

## International Story Competition 2017 – Thalassemia International Federation

### Story title – BLOOD RED by Dr. Namitha A Kumar

If someone were to ask me which color defines you best? I would instantly say “blood red!” One might think why this color? Isn’t it a violent color? Doesn’t it remind of you blood spilt? You would change your mind if you ask a child/adult with Thalassemia about the color red. It defines us as it defines me as it always did. The color of life! The color that puts back the color on my pale cheeks and white palms as the elixir of life drips its way into my veins. The color that ensures I will live. I will live to tell my tale many times over again. The color that stands for my life. A symbol of my life. A symbol of the blood bag and its magic potion that I receive every month. This is what keeps me alive. Blood Red...

My parents were expecting their first child and my family a grandchild! There was hope, excitement and enthusiasm when I entered the world. I had blue eyes at birth. A celebrity no less! Little did anyone even suspect that the Greek ancestry (beginning with Alexander the Great’s entry into India through Kashmir) flowing in my blood would play a game of delta and beta. A game of dice. Luck, fortune. Bad luck, misfortune. The die was cast. I was celebrated with great enthusiasm. Made much of by aunts and grandparents and everyone else. I had it all. The best of clothes, baby toys and lots of love. I seemed “normal.” I spoke at the age of eight months to much applause and pride. I walked at the age of one and a half and was treated to all the sweets available. I grew fat and plump. Baby arms and baby legs were played with by doting aunts and uncles and grandparents. I thrived till about the age of three. Suddenly I plummeted. Rounds of respiratory infections, fevers and bouts of vomiting left me pale and listless. I waxed and waned. I was taken to a round of doctors and hospitals. Vials of blood drawn from me and my parents. Anxiety mounted with impending test results. I was pronounced to have delta-beta beta Thalassemia or intermedia. I needed blood transfusions all my life to survive plus iron chelation and a range of medications and therapies just to hang on to life.

Life became a maze of doctors and a buzz of hospitals. Except my doctor aunt and my professor grandfather nobody in my family had ever heard of “thalassemia.” It seemed like an enemy. A snatcher of babies and a destroyer of childhoods. Enough! There will be no more babies in this family! How does one hold back tears when a needle enters a child’s vein and the child is screaming with pain? I was nearly three and a half when I got my first transfusion. I yelled, screamed and kicked. I was pacified with toys and treats. I screamed each time but was silenced with bribes. I came to associate blood transfusions with toys and treats. I hated it all! Not the gummy bear comics or the chucky duck or the black forest pastries could stop my screaming and tears. I would trade it for a normal child’s life.

I was taken to see Dr Bernadette Modell at the London School of Medicine. She gave us hope. I would live with proper treatment. I was taken to Switzerland at the age of five for a splenectomy. For the first time I met other children like me at CHU, Lausanne. The Thalassemia children’s ward was bright, open and spacious with two other kids. Deidre was seven and baby Lara was just two. I felt sad for baby Lara seeing her blood bag. I made friends with Deidre who immediately became my big sister! She showed me how to smuggle sugar cubes for baby Lara and get us strawberry yogurt from the pantry. I heard the docs

discussing something called desferal for the first time. I was scared. Deidre whispered to me, "if you keep quiet and don't cry much they will allow you to play in the nursery after dinner." I nodded. Dr Beck told me they would put a needle with a pump in my stomach. I was afraid. He was kind and I tried not to cry too much. Deidre was there watching. I was taped up after the poke and a pump was put into a sling with a pouch. I was told to go and play in the nursery. I did just that with Deidre at the lead. I saw the Deidre also had the sling and pump. I asked about it. She told me we needed it every night. How horrible! While playing with Lego blocks, I asked Deidre how come other children don't have IV poles and pumps and blood bags. Deidre the wise said they were "normal." I was not sure what that meant and asked her what was normal. She said normal kids didn't need IVs and blood bags. I understood vaguely.

After the nightly pump business, I was taken to another hospital in Moutier for an operation. I didn't like leaving Deidre and baby Lara and cried. This time the pediatric surgery ward had children a little older. Julian was 14, Erik was 13 and Lucia was 12. I was left alone in the ward at night with these children. I cried in fear. The other children came to my bed and consoled me. They taught me French words in the dead of the night and promised to show me IV pole racing in the morning. I was sold on it. The next morning as promised, we wheeled ourselves quietly to the nursery. There I was shown IV pole racing. Almost like a children's scooter, we pedaled the ground with one foot and raced on the pole with the other. Lucia screamed "just don't get the needle off your hands or you will be poked again." I raced faster than the rest and I was only a kid. I laid wagers with my small toys (a pink rabbit with long ears, a little guitar that sang Twinkle Twinkle and a glass globe with falling snow). I won more than I lost. I snatched 3D stickers from Lucia with bright sun splashes and watermelons; I got Julian's hotwheels key car and Erik's Spiderman toy! I counted my riches each night. The kids taught me French steadily and also how to play the xylophone. By then doctors were talking about an operation for me...

I didn't even know what an operation was until I came out of it. I yelled in pain and terror in the ICU as I spied the tubes and tanks and what not. There was a limp doll lying beside me. I threw the doll out...The splenectomy reduced my need for blood transfusions. I went in once in 2-3 months. I had stopped screaming by then. I had seen through the falsity that every child needs blood. I was the only one. That one specimen. I went to school and mingled in the crowds. Made friends. People at school knew I was different. I was not allowed to play much. My games teachers tried their best to interest me in chess. I insisted on running on the hot playground. I fell, I bled. There was confusion and panic. I was not allowed to bleed. I was sent to emergencies for a simple bleed. I came back to class with bandages and showed off! I was not given ice creams like other children. I was not given snacks (like sandwiches) that others got. Instead the teachers gave me chocolates and biscuits. I got my ice creams from other children on the sly. I was not allowed to drink the school filter water. The teachers kept my boiled water separate. I went and drank the tap water. I was the only character that was allowed to go home by afternoon. I did not stay till 3pm like the other children. I hated that. Other children envied me. They wanted to come home with me. I wanted to stay back in class with them.

Sometimes childhood was fun when the ugly serpent did not raise its hood. I was like any other child. I went with my cousins and family on trips. I climbed hills. I swam in rivers. I

played volley ball on the beaches. I trekked a hiking trail, rode a bike and did everything my cousins and friends did. Sometimes I got tired. Sometimes I was fine. My cousins and friends kept a vigil for the ugly serpent. I was checked for fever and rashes after each adventure. Sometimes I caught it, sometimes I escaped. On the no-escape days I was bundled straight to the hospital where doctors with koala bears on their stethoscopes checked me out kindly. I responded with venom and lashed out when they poked me. My cousins would entertain me while IV antibiotics or blood or whatever other medication was given. The hospital room was a stage while they did the chicken dance or whatever other stunt. One cousin would turn cartwheels all the way. My aunts would distract me from getting angry while my grandmother kept massaging my hand when I screamed that the IVs were hurting me.

I survived. I came back. I went back to school to tell my tales to my dazed classmates. I wanted to be a strong kid. I made friends with all the rowdy boys in my neighbourhood. They taught me to climb trees, aim with a catapult, make a bow with tree branches and use penknives. I played marbles on the street. I flew a kite on the terrace. I biked with them to the nearby lake to throw stones in the water. The boy gangs told me it was fine to eat ice cream anywhere. I did exactly that. Never mind my sore throats and coughs. I survived the worst. I was given vaccines each year. I hunted with penknives in my dreams...who I hunted I could never make out. It was probably the fire breathing dragon.

I had fun in school like any other kid. I made it to college. I was just like any other college kid. Like school, people in college knew. I was mad at my grandfather for telling the professors and the principal. Why should they know? My grandfather insisted they know in case I fell sick. And I never fell sick in those seven years. It was the best days of my life. The serpent was sleeping. I had grown stronger and outgrown all my childhood ills. I grew overconfident and rebelled. I insisted on using the desferal pump on my own. Most of the times I avoided doing it altogether without anyone knowing. A mistake I keep paying for till today...

My friends in college knew about the serpent. I told them casually one day. They were dumbstruck and shocked that I looked "like anybody else" and how could this be? They rallied around and helped wherever they could. If I had to go for transfusion, they would cover up for me and Xerox the college notes and record lectures. If I was sick and missed something important they would come home and let me in on whatever I missed. Once I had a viral and had to miss the college cultural show and my friends got their personal camcorders and recorded the whole show. We saw the show together in my house and my friends insisted on recreating a college show atmosphere with coloured screens and lights and smoke. The whole class turned up to watch the show with me. Lakshmi, Shilpa, Suma, Krupa, Anitha, Deepa, Meenakshi, Divyashree, Roopa, Ravi, Prashanth, Dan, Nitesh, Ramesh...my college friends whodid whatever they could to keep me happy.

The good of my being was every single friend of mine got tested for Thalassemia minor in college itself. Every single friend donated blood at the college blood donation camps. They joked, "Hey Namitha, this one is for you. Will you get my blood bag at your next transfusion?" I would retort, "Like hell I will get your blood bag! How many can I take? I just need one each month." There would be screaming, "Then ask for mine. I am also B positive!" I couldn't ask for better friends... or teachers. Teachers who took the trouble to

read about Thalassemia and educate their classes on the importance of carrier testing. Change begins at the school and college level. I would often see posters during the blood donation camps on free carrier testing for Thalassemia. I knew it was for Namitha...so that no more Namitha's would suffer...

True to my original Greek ancestry I loved Greek drama. English Literature was one of my core subjects and I thrived in its perfection. It was the perfect escape I had always imagined. Along with my friends I signed up for performing one of the trilogies of *Oresteia* (a play by Aeschylus). I imagined my family was cursed like the House of Atreus. Our drama professor Christine coached us on acting skills. I did a natural job there. My friends thrilled and remarked, "Like the perfect Greek warrior!" I retorted, "Greek tragedy is more like it!" My best friend Suma snapped I would be like Socrates and consume hemlock if I didn't watch my darkening hands. If Socrates died of hemlock I would die of iron poisoning. She knew of my playing hooky from doing desferal. The next weekend I tested for serum ferritin and it was a whopping 10300 when at max it should be within 1000 for someone like me! I was shocked out of my wits and knew I was done for. I knew it then I would pay a heavy price for this carelessness. I quickly got my act and started desferal regularly. By then it was too late...the damage had been done. It threw my endocrine system out of balance and I was left like a spinning top with no end...I would realize this years later when it actually hit really hard.

By the time I entered the MA English course in the fall of 2002, the Greek tragedy in my body had already begun. Only I didn't know it as I was asymptomatic at that time. I was healthy and moreover happy that I had begun a new course. I reveled in my talents for spinning out words, playing with language and deciphering complex theories. Encouraged and inspired by my professors I strived to be the best. I was fully into the texts. They became my life as did my friends from that class – Ashwini, Harish, Abdul, Shaista, Sarala, Roopa, Shilpa and Krupa again from my undergrad college. I forgot my troubles with desferal and the serpent and kept busy by escaping into absurdism, surrealism, modernism and post-modernism. Was it a coincidence I chose the part of Vladimir in *Waiting for Godot*? Would I be that Vladimir and my Godot was the ever-elusive gene therapy? By this time, there was deep and real research by Dr Michael Sadelain at Memorial Sloan Kettering on a very viable gene therapy cure. My grandfather had already met him and was full of hope that I would be cured of the serpent. He had been writing and meeting with gene researchers all his life and he had received hope from researchers like Dr Richard Mulligan from the Whitehead Institute and Dr Martin Cline from UCLA.

Meanwhile, I had totally detached myself from the serpent and ignored its ugly head. I refused to acknowledge it altogether and pinned my hopes on other dreams. I would top the Masters course and that was my hope. Nobody would be as proud as my professors and friends as I walked down the aisle at the university convocation to receive my gold medals for topping the course and standing first for the university. It was 2004 and the serpent was preparing a fertile ground for attack. I had forgotten to read the signs of the serpent.

Bang in the middle of my happy career, the serpent struck with such force and vengeance that was unimaginable. A fairly non-impact fall resulted in cracking the neck of my femur in 2006. People were shocked. It had to be screwed in place and this time I very well knew

what a surgery was! I lost my voice and mind...the only thing I recall is the title of Chinua Achebe's fiction *Things Fall Apart*.

"Turning and turning in the widening gyre  
The falcon cannot hear the falconer;  
Things fall apart; the centre cannot hold;  
Mere anarchy is loosed upon the world,  
The blood-dimmed tide is loosed, and everywhere"  
(From Yeats' poem *The Second Coming*)

In a flash, my happy life was plunged into hell as the serpent delivered blow after blow claiming revenge for every year of rebellion and carelessness. My liver enzymes were elevated indicating serious hemochromatosis and scarring. Ferriscans revealed blocked ovaries and pancreas full of iron and a wrecked reproductive system. I was lost in the maze of medical mysteries. Where would we even begin to correct this mess? This time, the docs came to my rescue as they not only delivered the right treatment but also got me back to the driving seat of life with their optimism and positive words. My best friends were my orthopedic surgeons Dr Shankar Kurpad, Dr Sunil KS and my endocrinologist Dr Arpan Bhattacharya. As usual Sameena was there to support with her expertise in hematology and transfusion medicine.

Slowly I got back to the life I had left behind but I knew life would never be the same again. Things had changed so much it was impossible to sweep up the ugly truths under the carpet. Then I discovered a community, another family. That was ThalPal, the online group. I recovered a little with Andy Battaglia's guidance and Sajid Rauf's prodding. Then there was Veeresh Pavate from Canada. His family had known my grandfather and had given us a lot of expertise in thalassemia. Again he taught me what to do and what not to do and how to manage endocrine complications. I managed to follow whatever people who been down that lane told me and I survived optimally. I regularly read up on TIF newsletters and updates. By then I had to take control and tame the serpent a little if at least to survive. I had arrived or rather had been thrown into the world of Thalassemia forcibly and forced to confront what I never wanted to. People told me it was time to stop pretending and accept things as it were. A complex issue of acceptance is best left alone.

Life with the serpent progressed as I enrolled for a PhD program in psychology. Amidst some more medical dramas and near-death I managed to complete the program. The serpent had dulled my senses and I did not get the same taste of victory on the day of my final defense of the thesis as I did on the day I received my Master's degree. Still according to everybody else it was a remarkable victory. I let others enjoy the experience. "One must imagine Sisyphus happy" (last line from Albert Camus' *The Myth of Sisyphus*).

My work for Thalassemia led to my professional work with other rare diseases as I discovered we are not alone and sometimes felt fortunate at least we have treatment protocols and available drugs. When I look at the record of the children who died of Pompe disease or other lysosomal storage disorders before the enzyme became available in 2001, I feel at least I was born at a time when protocols for Thalassemia were already well-documented. When I see children with rare disorders like Niemann-Pick syndrome or Rett's

syndrome or Fabry's disease I understand that Thalassemia is not as bad. We know there is a path ahead if we chose to go sensibly not like what I did rebelling with treatment. It is too late to turn the clock backwards but I always wish I could. I have to live with an impaired endocrine system maybe until the time gene therapy reverses it (a lasting hope!).

Sometimes I hear the dim roar of the blood red tide crashing in my head as I face more medical challenges and dramas with Thalassemia. Bag after bag of blood red is not enough to tide the lack as each crisis brings in hemolysis and plunging hemoglobin levels. Nothing stays, nothing is still. Everything breaks down, falls apart only to rejoin and come together again. We live if only with hundred and one scars and innumerable scares! Still we live to tell our tales again and again...stories of blood red, the gods of Erythro and the goddess of Thalassa and Alexander's army marching into India and the throw of dice of delta and beta and alpha.

Still among Pandora's Box of troubles there is hope...

"Surely some revelation is at hand;  
Surely the Second Coming is at hand."  
(From Yeats' poem *The Second Coming*)

Surely the "Second Coming" is at hand; surely we live in hope of getting gene therapy. Every single one of us lives in the hope of being cured and getting a second life. Blood red will be put to a final rest somewhere in the future perfect.

"None but the brave,  
None but the brave,  
None but the brave deserves the fair."  
(From Dryden's poem *Alexanders Feast*)