

Personal History: A Myeloproliferative Disorder in Pregnancy

Two years ago, when I found out I was pregnant, I didn't know what a myeloproliferative disorder was.

My husband John and I were thrilled to find out we were having a baby. We had had trouble conceiving a child, and we decided to try IVF. Our odds of success were very poor – only 10% or 15% - so our doctor recommended that we try a very high dose of hormone stimulation. I had an operation, and once I healed, I started with hormone stimulation. Every day John gave me four injections with inch and a half needles. All in all, I underwent two operations, spent \$17,000, had 100 intra-muscular injections, and missed an awful lot of meetings at the office.

The night after the embryo transfer, John and I sat at the table eating dinner. "Just a minute," he said. He disappeared into the living room and returned with two candles and a framed picture. The picture showed an electron microscope photo of our single surviving embryo, eight little cells forming a mysterious looking little ball. John set the picture on the table, and surrounded it with candles. "Think good thoughts," he said, if you get pregnant," he said, "maybe you should quit your job in San Francisco and we'll go to London."

A week later we got up early on a Saturday morning for my blood test appointment. John handed me a cup of coffee, which overwhelmed me with an emetic smell. "Get that away from me," I said, taking a few steps back from the cup he was extending in my direction.

Our infertility doctor was thrilled when I said I was nauseous. "The sicker you feel, the better I feel," she said. She was even more pleased with the blood test results, and we found out that despite the odds, I was pregnant.

Five weeks later, I saw the embryo's heart beating on the ultra-sound, a tiny "equals" sign flashing on and off. "You're graduated," said my infertility doctor, "low chance of miscarriage. Go find yourself a good obstetrician."

The next week I went for my first appointment with my midwife at Stanford Hospital. The waiting room was filled with pregnant women of every nationality, some holding their partner's hand or with toddlers in tow. After the anxieties of the infertility clinic, where weeping couples poured over statistics and whispered to the financial advisor, it was a cheerful scene.

I beamed as the midwife gave me piles of brochures to read, and happily extended my arm for a blood test. The worst was over, I was sure.

The next day I had a phone call at work from the obstetrician. "We want you to come in and repeat your blood test," she said. "We had some worrying results."

"I had IVF," I said. "I had two surgeries before I got pregnant. I went through a really intense protocol."

"Your platelets are 770," she said. "We think we might have made a mistake you're your test. But we're concerned..."

I tried to remember anything I might have learned about platelets back in high school biology. They were shaped like discs, they made blood clot...

"We're concerned you might develop a DVT or even a stroke. Can you leave work now and come in for a test?" said the obstetrician.

Over the next few weeks, I learned more about platelets. The obstetrician referred me to a

hematologist the next day. I went back to the hospital, this time not in the cheerful midwife's office, but in the cancer center. Young kids, maybe 16 or 17 years old, wearing breathing apparatuses over their faces, played video games in the waiting room. They looked like pale, sickly, young Darth Vaders.

The hematologist was puzzled. Had I had a bad reaction to the IVF drugs? or an infection? She sent me for more blood tests. I went for an ultrasound to look at my spleen, which turned out to be enlarged.

The day I came back for the results the hematologist was very busy and I waited about an hour to see the resident. It was cold in the examination room, and I wrapped myself in a sweater and took sips from a water bottle as I waited, starting to feel pregnant. I looked at copies of Blood Magazine stacked on shelves.

The resident came in, a slightly overweight young man with a self-assured, breezy manner. He sat down across from me at the desk and sneaked a surreptitious look at my just slightly visible bump.

"As it turns out, the IVF had nothing to do with your platelets. You had this platelet problem before you started with the IVF," he said. "So, let's start with the good news," he continued, bouncing a little in his seat. "The good news is that we tested you for the Philadelphia chromosome, and you don't have CML." Then he paused, looking at me. "What you've got is a myeloproliferative disorder." He took a breath. "Now, this is a whole range of diseases, a continuum, going from the very benign...(pause)...to the very serious." He gave the desk a quick karate chop on the left. "Very benign here," he said, then chop chop chopping with his hand, across the table left to right, "very serious there. This is a range of diseases that begin with the very manageable and moves up to CML. But you don't have that. You haven't got leukemia. Although we've got great treatments for leukemia now. We've got Gleevac. Bone marrow transplants are much easier than they used to be. But you haven't got leukemia."

"Okay..." I said, "But what have I got? Where do I fall on this line? And what about the baby? Is he okay?" I didn't feel panicked, but confused by this information. All the table chopping meant what? Which chop was my disease? I thought of a colleague of mine, who had died of leukemia two years after surviving a bone marrow transplant.

"We can't give you an exact diagnosis without a biopsy, and the results wouldn't change the medication plan anyway. You don't need the biopsy right now, you can have that after the baby is born. You should take one low-dose aspirin per day and work with your obstetrician."

I went home and reread the report my hematologist had written to the obstetrician: "Patient is alert, friendly, and in no obvious discomfort. Patient has enlarged spleen, platelets over 700. I viewed at her slide myself and saw occasional tear-drop shaped red blood cells. I have not ruled out the possibility of a myeloproliferative disorder, which would most likely be early myelofibrosis."

I didn't know what this meant, but it sounded nasty. I surfed the web, looking for myelofibrosis and pictures of red blood cells. Most of the websites indicated I would be dead in five years. That night I lay awake at night, thinking about fibrous matter inside my bones and strange, drop-shaped blood cells and platelets clogging my veins.

A month after I had the hematologist's report, I was five months pregnant, and John and I had packed up our entire flat and we were on a plane headed for our new life in London.

Our first order of business in the U.K. was to find a hematologist and obstetrician, and so we started on a new round of visits to the doctor. I waited several weeks for an appointment with the hematology unit at our local hospital. In the meantime, my platelets came down to about 550 as my blood volume increased with pregnancy. My blood pressure started to creep up. The

midwife would take my B.P. every two weeks, look questioningly at the gauge, then send me out for a cup of tea to relax, and take my pressure again. Sometimes it hovered around a higher number, sometimes lower. "Maybe you're just anxious having your blood pressure checked here in the hospital," she said.

My new hematologist thought it would be best to proceed with the biopsy. There was a few weeks' wait, and by the time the date rolled around, I was seven months pregnant. I laid on my side for the biopsy, with my seven-month belly extending out to the right. The doctor stood behind me, digging with his needle to get into my hipbone. He muttered under his breath as he ground the needle in. I touched the baby as he continued to dig. "It's okay," I told the baby. "Just sit tight. You can hear it but it won't hurt you." Baby (now code-named Andrew) kicked hard.

After what seemed like 15 minutes or so of these maneuvers, Dr. M. gave a sigh and took the needle out. "I am having trouble getting marrow," he said. "The needle is bent." He showed me the needle, with its tip bent over and blunted, apparently on my hipbone. He shook a little tube with bloody liquid in it, and a piece of bone. "It could be that I couldn't get the marrow because the needle was bent. Or it could be that there wasn't much marrow."

The nurse came in with some tea. Dr. M. walked out of the procedure room, and spoke to John in the hallway. He spoke softly, possibly to avoid my overhearing him, but I could still make out what he was saying. "Sometimes people get bad news," he said to John. "Sometimes you are quite young, although your wife is forty, that's not really so young, but you are young and you think everything will go well and...sometimes people get bad news."

I woke up feeling frenzied at 2 a.m. "What if I die?" I said to John. "Who will help you with Andrew?"

"You won't die," said John.

"Everybody dies," I said.

"You won't die now," said John.

"Maybe I'll be dead in five years. Maybe I don't have any bone marrow."

"Look, try to get some sleep. It can't help to stay up all night."

"I need you to promise to take Andrew to see my parents."

"I promise."

"You also have to promise to keep the house clean. I don't want to look down from heaven at you and my son buried under piles of newspapers and books and dust."

"He'll like to read newspapers."

"Do you promise to keep the house in order?"

"I promise," said John.

"Look, hire a cleaner in to help you," I pleaded.

"I'll check into the cost," he said, "it could be expensive."

"So if I'm dead you're going to worry about the cost?"

We were silent for a few minutes.

"Anyway," said John, the most likely scenario is your doctor had problems doing the biopsy. He probably bent the needle and didn't get any marrow. It could happen."

"So I should relax because he might not know what he's doing?"

"Let's get some sleep," said John.

"He said forty was old," I said.

"See, he can be wrong about things," said John. "Forty is the new thirty."

I had only been asleep for a few minutes when John woke me up poking my shoulder with his finger. "Roll over onto your left," he said, "it improves circulation. Left - think Left."

I didn't have results from the biopsy for nearly a month, but what I learned was anti-climatic

compared to what I had feared. I probably had essential thrombocytosis. I should keep taking low-dose aspirin and have a heparin shot or two after the delivery. No big deal.

Even so, I became desperate to switch to another hospital. Aspirin didn't seem like much of a plan. I thought about going to the U.S. to have the baby, but the larger I got the more dangerous that seemed. Although the hematologist was not excessively concerned about my delivery, as the baby got bigger it made sense to me to switch to a hospital with more specialists.

In the end, I worked with my G.P. to change hospitals to St. Thomas'. I was already nearly eight months pregnant at my very first pre-natal appointment there. The midwife saw me late in the day, at six in the afternoon when the clinic was already empty. She talked to me for a long time, taking my history, and just as we were finishing she took my blood pressure.

"Let's take this again," she said, looking at the reading. She glanced at the telephone. "Let's see if the registrar can talk with you." She made a few phone calls, and I glanced at my watch. "You might want to call your husband," she said, "I don't think you're going home."

I called John and he came down to the hospital. All the night units were full, so eventually they moved us to a delivery room. We were too late for dinner, so we ordered some take-away Chinese noodles. We listened to other women hollering and screaming, delivering their babies, as we looked out at the London Eye and the lights over the Thames, and ate our noodles out of boxes with chopsticks.

My blood pressure was very high when I went into the hospital, and my platelets were about 750. The registrar decided I should stay so they could watch me for pre-eclampsia. I remained in the hospital for a week while they monitored me and checked for protein in the urine. The platelets were potentially blocking blood flow into the placenta, causing my blood pressure to rise.

After a week in the hospital I was dying to go home. I didn't have time to shuffle around the television room in my slippers and my pressure tights, watching the Iraqi war on the news. I wasn't prepared for the baby yet, no nappies, no baby clothes, no sterilizer, all my work at the office left up in the air...I sat in bed in my pajamas, making lists of everything I still had to do: buy baby suppliers, paint the dining room, make curtains for the baby's room.

After a week of collecting my urine in a giant bottle, the obstetrician told me I had no protein in my urine, so I could go home. I needed to have my blood pressure checked every other day, but I could stay home. I was thrilled at the prospect of getting out of the hospital, into the fresh air and home. It was the end of March and spring was around the corner. I was eight months pregnant and still had a month to go before the baby was due.

Before I left the hospital, I met with a new hematologist from St. Thomas'. This time the discussion was more worrying. I had some red blood cell involvement, they thought, and I might very well have P.V. My blood pressure was climbing from platelets blocking blood flow in the placenta. The hematologist discussed my case with the obstetrician, and this time, to my relief, rather than writing a letter they spoke on the phone.

Although I had spent my entire pregnancy frantically worrying, when I left the hospital I put it all to the back of my mind. For the next two days I rushed to MotherCare, Boots, John Lewis and all over town, shopping for baby gear. I made an appointment to have my hair cut and to get a manicure. I bought special baby towels with hoods and a baby cardigan with caterpillars embroidered on it. I collected bags and stacked them into the car. Late in the afternoon, after my shopping, I felt great, and I wondered if it was really worth going all the way back to the hospital to get my pressure checked. The baby seemed quiet, though, he wasn't kicking much, although maybe I hadn't counted his movements that well, since I had been running around.

I stopped by at my local midwife's clinic. She was annoyed. "You changed hospitals and went to

St. Thomas’,” she said. “We can’t treat you here now.”
“I just want you to take my blood pressure,” I said. “If it’s high I’ll go back to St. Thomas’.”
“I can’t take you as a patient anymore,” she said.
“Just a quick check. Just my B.P.”

She slipped the blood pressure cuff on my, shooting darts. She listened for a minute, paused, and then said, “Let’s run a quick scan while you’re here.” The baby’s movements on the scan seemed sluggish. I started to feel my breath tighten.

The midwife called the registrar. “If he gives you the okay,” she said, “you need to go to St. Thomas’ now.”

This wasn’t my original plan. We had a hotel room booked near the hospital for around the delivery time, but we were a month early. The registrar came in and looked at the scan. “How long will it take you to get to St. Thomas’?” he said. “An hour,” I said. “Go down right now,” he said. “You’ll be fine.”

I called John. “Can you meet me at the train station?” I said. “We can take a cab from there.”
“It’s pretty fast to take the tube,” he said. “Why take a cab? It’s not that fast in the traffic anyway.”

I took a deep breath.
“Listen hunny,” I said. “Get yourself down here right now.”

That night me and my bump were back in the hospital. I looked at the tape every minute to see if how Andrew looked on the scan. He seemed quiet and more droopy than normal, not the usual jumpy up-and-down spikes on the scan.

The obstetrician decided to induce me that night, due to my ‘dodgy’ blood pressure. John waited until midnight, and in the end he fell asleep on some cushions on the floor. When we woke up in the morning, we waited for the midwife to start inducing me, but the staff was busy and we didn’t get started right away. I thought about all the platelets running through my arteries, and how much they were clogging the placenta. Was Andrew getting enough oxygen to the brain? Was his heart beating effectively? At nine A.M. the midwife finally came in with some gel to start inducing me. It was going to take 12 hours or even 24 to deliver the baby, she said. She and the doctor were happy with the scan, and things seemed to be going all right. I relaxed a bit.

I thought of my nice suitcase back home, full of delivery room kit, with my copies of the New Yorker magazine, my CD player and my massage oil. I wanted some distractions. “Maybe you can go home and get my suitcase,” I said to John. “I think we’ve got time. You could have a shower and a shave.” We debated, but 24 hours seemed like a long time. In the end John headed home to get my gear.

I turned back to looking at the scan while I waited for the gel to have an effect. After an hour or so, I felt a first, tiny, almost imperceptible contraction. Andrew’s heartbeat fluttered on the scan. It slowed for a long minute and then came back. At my second, tiny contraction, his heartbeat dipped and then there was a long, long gap. I watched the tape, waiting for the heartbeat to come back, my own heart racing. I started to fiddle with the scanner belt to try to get it into the right location across my tummy. The midwife saw me and came back into the room “I think the belt might be in the wrong place,” I said with some panic in my voice. We both looked at the tape. Oh, Oh, Oh. Dr. D. isn’t going to like this,” she said, and disappeared out of the room.

A minute later, four or five nurses and doctors came in. Dr. D., a tiny, sharp woman, looked at the scan. “This baby has decided not to wait,” she said. She turned on her heel and looked back at her team. “Let’s get her into the operating room.”

“My husband isn’t here yet,” I said, panicking. “He’ll be back in 30 minutes.”

"He'll have a chance to meet the baby then."

My eyes were welling up with tears.

"No crying," she said sternly. "I don't want crying in my operating room."

I called John on my cell phone as they started to wheel me into the operating room. "I can't find your pajamas," he said. "I thought you said they were in your suitcase." "Forget it," I said, "Get back in the car and come back. I'm having a caesarian. The baby will be here in 30 minutes." I rang off as we spun around the corner into the operating room. One young woman was trying to start an IV into my hand. She fiddled around with the needle, feeling for a vein, and taking a long time. "Give me that," said someone tall behind me, who turned out to be the anesthesiologist. He grabbed my left arm and inserted the line in less a second. I suddenly felt I would be extremely grateful for all my life to someone who could solve a problem in a split-second. A curtain went up below my shoulders, and I could hear metal objects clanking around in a tray. "Can you feel this?" said the anesthesiologist, pinching my arm. "I can feel that." "And can you feel this?" "I'm not sure," I said slowly, but as I was speaking he nodded to the people behind the blue plastic curtain and I felt an odd pulling sensation.

"Hurry and get the baby out of there," I said to the anesthesiologist, tears running down my temples. If he could breath himself, I thought, he wouldn't be dependent on my platelet-clogged placenta for oxygen.

"Everything's fine, you're doing really well," said one of the nurses, holding my hand. I heard a tiny meow-like chirp sound, the sound of a tiny cat.

"Is that the baby?" I said. "Is he okay?"

"He's okay," said the nurse.

They didn't bring him over right way, and I asked the nurse again, "What are they doing? Is he all right?"

One of the other nurses showed me a little bundle of blanket, with a very tiny brown and wrinkled face, like a little brown apple with squinty eyes. The nurse held him out to me to hold as I lay on the table, but I refused. "I can't hold him," I said. She looked hurt and surprised. "Please check him again. Please make sure he's okay. Take him back and make sure he's okay." "He's fine," she said.

Just then John burst into the room through the swinging operating room doors, in a blue gown and cap, clutching his camera in his hand. He gave me a kiss, then ran to see Andrew and give him his first bath. A few moments later, back in the delivery room, we were cuddling our baby together at last.

Andrew was a small baby, five pounds fourteen ounces, but lively and adorable. I stayed up all night watching him in his clear plastic bassinet, checking to make sure he was still there. I stayed in the hospital for a week, and the day I was released I was (again) thrilled to go home. As I signed the paperwork to be released from the hospital, one of the midwives leaned over the desk to me. "I saw your baby's blood test results from right after the delivery," she said. "That was close, wasn't it? It was awfully close." I couldn't even answer – I didn't want to know how close it could have been.

John and I packed little Andrew into his car seat. He seemed like a very tiny space creature, lost deep in the cushion. We both felt happier than we had ever felt before, each of us holding the handle on the car seat, carrying our baby together. Outside it was a gorgeous, bright, sunny spring day. The tulips were in bloom in St. James' Park as we drove past Buckingham Palace.

Back at home, I walked up the stairs to take Andrew to his room and settle him in his Moses basket by the window. He seemed very cozy snoozing on his fresh linens, with all his toys and

his little outfits nicely laid out in his chest of drawers.

As I turned to come downstairs, I felt something sticky on my dress. Did I get water on it washing my hands, I wondered? I looked down and saw a puddle of blood on the floor. I lifted my dress to see a whoosh of blood shooting out of my caesarian wound, spraying like water from a hose. We headed back to the hospital.

As it turned out, we could blame a lot of this on the platelets. I was taking heparin injections to avoid a blood clot, and although I had a lot of platelets, they were pretty low-efficiency, malfunctioning platelets, so many rogue traders using shoddy building materials. After a few days I stopped losing so much blood from the wound, but I continued to bleed for a long time, and a nurse came to the house every day to dress my wound for nearly 18 weeks.

With Andrew safely delivered, the frightening part of the story seemed to come to an end. It was a risky time for me, though, as my platelets shot up to 1.3 million in the two weeks following the delivery. I had lost some blood during the caesarian and felt weak. Andrew was small and needed to feed every two hours, so we did not get any rest for many weeks. It took me a while to regain my strength, but slowly Andrew got bigger, my platelet count came down, and I felt better.

Today Andrew is 16 months old, running up the stairs, saying "Hello Duck!" to the pigeons in the park, and up-ending his milk bottle on the carpet. John has not had to worry about the cleaning bill, because I pay it without telling him. My platelets are about 700 and I'm taking a low-dose aspirin every day. We are lucky and we are all glad to feel healthy and alive. We're even thinking about Baby No. 2, this time with more heparin.

I would like to thank all the staff at St. Thomas', the Hematology department, my midwife, and the nurses and doctors who delivered my baby safely and protected my own health. I am extremely grateful to the NHS for all the excellent care and follow-up that they provided.

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