

## Activity 3.1.2: Diary Entries

### Anna, age 10

So, I'm back in the stupid hospital again. This time, the pain is in my chest and my leg. My chest pain is really deep, making it hard to breathe—probably an 8 on that 1 to 10 scale the doctors always ask me about. And my leg is throbbing so much that it even hurts when my mom tries to rub it for me—it definitely gets a 10. I tried all things the doctors tell me to do at home—take my pills, take a warm bath, rest...but nothing worked and my mom had to bring me here. Plus I had a fever this time and everyone seems to get really nervous when I get a fever. I guess they get worried I have some sort of infection, which mom tells me can be really, really serious for someone like me—someone whose spleen doesn't work like it should (they showed me a picture of a spleen once—it was purple!). Until I turned 6, they actually made me take antibiotics every single day to try and keep me from getting infections. Now I don't have to take those pills anymore, but my mom makes me wash my hands like 50 times a day...

Anyway, we sat in the emergency room for like four hours. The nurse had to stick my arm FIVE times before she found a vein for my IV. The needles don't bother me too much anymore but I am SO bored right now. Some kids think it must be fun to sit here all day and watch TV but I'm sick of it. Plus they've got me on the pain meds that make me feel so sleepy so it's hard to even stay awake.

All my doctors and nurses are really nice, but I'm still really bummed because this week is the science fair at school and I worked really hard on my project. And how am I going to make up all the work I keep missing? I used to get straight As, but now it's really hard to do that cause I miss so much school for doctors appointments and trips to the hospital. I get a little sick to my stomach when I think about all the work I'm gonna have to make up this week. I wonder sometimes if Mrs. Collins really gets why I'm out so much—I don't want her to think I'm a bad kid, or a stupid kid, or a kid who pretends to be sick to get out of going to school.

Sometimes I really hate this sickle cell thing. It's always getting in the way!! I'm so sick of missing out on things at school and with my friends. Like last month? I didn't get to go to my friend's birthday party because I was in pain...It stinks! I miss out on a few days of school or a few activities and it's like people kinda forget about me. None of my friends have sickle cell disease so no one really understands. Sometimes I just feel like screaming! Ugh!

Well, I gotta go cause physical therapy is here to help me get out of bed and do some exercises. Usually, we walk up and down the halls of the hospital together to try and get my legs moving. It hurts at first but it always hurts worse if I just sit in bed all day. After we do the exercises, they let me sit in the warm whirlpool for a while, so I always have that to look forward to. And later, I have to get that ultrasound again...I think they call it a "Transcranial Doppler"...I can't remember what that one is for (I think maybe it checks to see if there are any problems with the blood in my brain) —there are so many tests I can't always keep track.

### **Anna, age 17**

This is my second month on hydroxyurea. The doctors decided it would be a good idea to start it since I've been having so many pain crises this past year. They said this medicine is pretty new but that it can actually help me live longer. It should make me have fewer pain episodes and keep me from getting acute chest syndrome again (that was NOT fun—I actually ended up in the intensive care unit for a few days because my sickle cells were blocking blood flow to my lungs and I was having a lot of trouble breathing). I'm not so sure yet. I had another crisis last month but they say it's still too early to tell if the hydroxyurea is gonna help me. All I know is that it makes me kinda nauseous and tired, which they say is pretty common. Oh, and I think it's making my hair fall out, which is NOT okay. Seriously, the other day I was combing my hair and I swear it was like I was shedding! My mom says you can't tell, but I'm totally afraid I'm gonna wake up one day and have a big noticeable bald spot. So not cute. I already have weird yellowish eyes from my sickle cell and now I'm gonna have thin hair, too? It just keeps getting better...

Oh! I forgot the worst part! The doctors said that if I were to get pregnant and have a baby when I'm taking hydroxyurea, there's a chance the baby would have crazy birth defects. Now, I'm not planning on having a baby anytime soon but they made me go on birth control just in case.

I'm trying to stay hopeful that this medicine will make things better not worse. I try to keep a good outlook, but sometimes it's really hard. I feel different, look different (at least I think I do), and can't do all the things my friends get to do. Just last week, my doctors told me that going on my senior skiing trip would be a really bad idea...you know, cause of the cold temperatures, the high altitude, and the bone problems in my hip. I begged and they agreed to let me go, but I have to stay in the lodge, bundled up the whole time...meaning I miss out on all the fun my friends will be having on the slopes. Most of my close friends know the deal with me by now and they try to be really supportive, but I can't expect them to sit back with me and stop living their lives. Sometimes my sickle cell just makes me feel really alone. I know adults always say that being a teenager is hard, but trust me, being a teenager with this disease is a whole different ballgame.

### **Anna, age 22**

I just don't know whether I should do it now or try to manage a little longer with this pain. My orthopedist says the avascular necrosis in my hip joint is just getting worse, which is why walking is becoming more and more challenging. Apparently, the last x-ray showed that the round ball part of my bone is all rough and jagged. The doctor says that basically, the bone is dying (causing my joints to collapse) because the main artery that supplies the blood to the bone has been blocked again and again by the sickle cells. I've been on daily arthritis medications for a few years now and always make it to my physical therapy appointments (which can be pretty torturous if I do say so myself). I even used a crutch last spring to try to give the joint some rest. But it still hurts. And so I'm faced with the decision about whether or not to proceed with a hip replacement. Yup, a hip replacement! You know, the surgery that 80-year-old men get after falling? That's what I'm up against right now.

My parents think I should do it, and I sort of understand the reasoning. I'm in chronic pain and it really takes a toll on my life. All the medications, physical limitations...sometimes I actually DO feel like an 80-year-old man. I'm not as mobile as I would like to be and it makes it hard for me to go out with my friends and enjoy life like

college seniors are supposed to. Plus, it's SO difficult getting around campus. I have to get up earlier than I'd like just to make sure I make it to class on time, and then when I do, I'm in pain and exhausted – kinda makes it hard to concentrate on Literary Theory when all you can focus on is the throbbing in your leg. Just last week, I talked with the Office of Disability Services and they said they can try to make sure my next semester courses are closer together or they can arrange for security to come take me to and from classes. I'm already that girl who walks with a limp—do I need to broadcast further my disability with a security chauffer?

My big worry is that the doctors have said that if they do my hip replacement now, I will need another one in about ten years. Yup, apparently these artificial hips are only made to last about 10 years (which wouldn't be a problem if I actually were an 80-yr-old man)...so I'd need another one in my thirties, forties, fifties...you get the picture. Plus it's a pretty major surgery—I'd at least wait until I graduate but then I'd still be looking at putting off the job hunt for several months...and I'd probably have to move back home for a while. Recovery and rehab would be a pretty long process. But maybe it would be worth it?? Who knows, but right now, with this constant pain, something's gotta give...

### **Anna, age 31**

Alex and I have been talking more and more about starting a family. Since I've been off the hydroxyurea for a few years now, there's no longer a greater risk for birth defects, which of course is a good thing. That being said, Alex has sickle cell trait (we found out just last month – he had no idea), which means that genetics are really not in our favor. I've always wanted to have kids, but thinking about the chances of having a child with sickle cell, knowing all that I've gone through – I can't really wrap my head around my own child having to deal with the pain, the hospitals, the complications...And I would be *knowingly* taking that chance? Would I end up with a constant feeling of guilt, feeling responsible for doing this to my child? Or could I be in a better place to help him or her through it all since I've been there myself? Why does sickle cell always have to make everything so complicated? I would really love it if just one decision in my life could be a simple one...The stress is really taking a toll on me and in the past few weeks, I've notice that it's really taking a toll on my relationship with Alex.

And then there's my mom. She's not so thrilled about the idea of me getting pregnant. She's worried about my health and the complications that could arise. Just the term "high risk" makes her get all frantic – and I suppose I can't blame her given all she's had to see me go through over the years. Every time she calls, she's worried about something new – whether the stress of pregnancy puts me at greater risk for stroke, how my artificial hip will manage the strain of pregnancy...she makes good points and I know she's just worried because she loves me, but sometimes I feel like she's not being supportive enough of my desire to have a family.

During my appointment last week, my hematologist recommended Alex and I meet with a genetic counselor. He gave me a recommendation of someone he works with over at the University Hospital, which generally has a great reputation. I suppose it wouldn't hurt to have someone who specializes in this sort of thing talk us through our options. I've done some research online (even though my docs always tell me not to!) and I've read that some couples decide to avoid pregnancy altogether (and start families other ways), some choose to have the baby only if prenatal tests show that the fetus is healthy, and some choose the help of preimplantation genetics. This last option was totally foreign to me, but after doing some research, I can't say I'm not intrigued.

The idea of being able to choose only those embryos that are genetically free of sickle cell disease? That I could be sure my baby would be healthy? Who knew the science existed for that sort of thing? It's really pretty amazing...but all still really scary.

I suppose I should just go ahead and make the appointment with the counselor to find out more. It can't hurt, right?