

Lessons on Sickle Cell Disease

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One spring morning during my intern year of residency, I arrived on the medicine ward to discover that the overnight resident had admitted a patient in sickle cell crisis to my panel. This patient was only 27 years old, and had already suffered ten episodes of acute chest syndrome, three prior strokes, and avascular necrosis of the right hip. These complications had left her severely debilitated. She was now living in a permanent nursing home and would visit our hospital every couple of months for management of sickle cell crisis. The overnight resident had suggested that I spend as little time with the patient as possible. The resident had placed her on a Dilaudid patient-controlled analgesia (PCA) pump, and the patient seemed comfortable when alone. However, whenever the patient sensed someone else in the room she would immediately writhe in pain and push the Dilaudid pump for meds. Better to avoid her and not cause undue stress to the patient or the hospital staff, the resident advised.

I quietly slipped into my patient's room. She was asleep and was without a grimace on her eyebrow. In keeping with standard protocol for every patient on my service, I gently shook her awake to see how she fared through the night. Slowly, she opened her unfocused eyes as if coming out of a pleasant midsummer night's dream. When her gaze focused onto me she immediately released an agonizing scream and begged me to make the pain stop. She reached for her Dilaudid pump and quickly pressed it for relief. Moments later the patient's nurse rushed into the room and tried to shoo me away. Overnight, the nursing team had spent an inordinate amount of time trying to gain IV access for the patient's Dilaudid pump. Therefore the nurse wasn't going to allow me to lose the precious access off the patient's pinky finger because I didn't know what I was doing. When we were both outside the patient's room, the nurse quietly echoed what my overnight resident originally suggested: that the patient seemed to be in pain only when someone was near her. My patient, in the nurse's opinion, was a drug seeker.

During morning rounds, we reviewed my patient's chart. Her bloodwork was consistent with hemolysis, and she was very likely in an acute crisis. The attending suggested that my patient might also be suffering from a concomitant pain syndrome in which patients perceive legitimate pain, but experience that pain as something more intense than what they are actually feeling. Regardless, the plan remained the same: pain management, IV fluids, and supplemental oxygenation. If needed, we could insert a PICC line for more secure IV access. We then agreed upon the specific settings for my patient's Dilaudid pump, and I was instructed not to deviate from (i.e., increase) this setting.

During the first couple of days of my patient's admission, I made an effort to check on her as much as possible. Without fail, she would wince in pain and press her Dilaudid pump every time she saw me. But during those brief moments, she also shared snips and pieces of herself: her former life as a college student; her current life as a nursing home resident; her general loneliness as the only person under the age of 70 at her nursing home; her idea to educate medical students about the experiences of living with sickle cell disease because "they just don't get it;" her relationship with her father. I came to enjoy those moments together.

Then one day, she lost IV access during the middle of a pain crisis. The nurses didn't have to page me because I could hear her down the hall. My patient begged me not to attempt to place another IV line because it would hurt too much. I didn't listen to her. I also didn't regain access. Her oxygenation saturation was fine, but she was tachycardic. I asked her nurse to inject a hefty dose of Dilaudid. The nurse asked if that was advisable since the dose that I suggested was greater than what the patient would normally receive over the course of an hour on the Dilaudid pump. Because my patient's morning labs demonstrated hemolysis, and it was unlikely that she was faking tachycardia, I reasoned that she must be in true pain, and not just drug seeking or experiencing a pain syndrome. However, the injection didn't relieve her. My heart sank.

I placed an order for a PICC line. I had nothing left to offer to my patient, so I sat by her bedside. To this day I do not really understand what I was trying to achieve, nor do I know what I actually achieved, if anything. But for ten minutes I sat at my patient's bedside and bore witness to her pain. We sat together in silence. I offered no words. She volunteered no words. She never asked for more medication. She simply cried, and screamed, and moaned. I reached for her hand, and she allowed me to take it. The nurse would come in regularly, but she would quickly leave once she saw that I was with my patient. At one point during those ten minutes a member of the housekeeping staff entered the room and mopped the floor without acknowledging either one of us. I wondered how often this moment reflected my patient's reality: to cry out in excruciating pain, only to realize that everyone who walks by doesn't recognize that she even exists. I had never previously contemplated the possibility of such abject isolation.

I told my patient I needed to leave to coordinate the insertion of her PICC line. I left her room, with her pain ever ringing in my ears and mind. By the afternoon we had regained IV access with a PICC line and provided my patient with proper pain medication. By the end of that day, I thought I had learned something critical about sickle cell disease and crisis.

Specifically, I thought I learned that the key to effective pain management was not the proper selection of opioid medication, but the cultivation of a meaningful human connection. More than five months have passed since I discharged my patient. She has not yet been readmitted, and I imagine that she is still in the nursing home, alone and lonely, without a chance to return to her former life. As I reflect on it now, months later, I believe I have learned only one thing about the pain of sickle cell disease: I simply don't get it.

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