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## HEALING ARTS: MATERIA MEDICA

# Living Fully with Sickle Cell Disease

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***Innocence.*** She is a 17-year-old African-American female born with sickle cell beta plus thalassemia. Through no fault of her own, she is genetically marked for life. Over her lifetime she has had several admissions for infections, three blood transfusions, and two episodes of acute chest. Pain due to vaso-occlusive episodes (VOE) is usually managed at home. The miracle of hydroxyurea, started at age 12, improved her symptoms significantly. Nearly 5 years pass with stable labs and no pain episodes. Almost forgetting that she has the disease, she is living a seemingly "normal" teenage life. She is an excellent student with a loving family and good friends. She enjoys playing the piano, listening to music, theater, and participated in high school gymnastics. Her disease is described as mild and ibuprofen is the pain medication she typically needs.

***Pain.*** Out of the blue, a few weeks before her 17th birthday, she has a pain episode, while relaxing at home with none of the expected triggers. In a matter of hours, she is in tears with severe pain. The pain is in her back. She misses school the next day, a Friday, so she has all weekend to recuperate. By Monday, she is well and back at school. In a way, this is good. In three weeks, she will travel on her own to Paris to visit her aunt; this is a refresher on how to handle VOE pain. The school year ends, and off to Paris she goes.

Just a week into her trip, another pain episode. The pain lingers; armed with her pain medications, she soldiers on, determined to enjoy her vacation. Eventually, she is admitted with a fever, chest pain, shortness of breath, and back pain. After an overnight stay, she is discharged. She is leaving to return to the USA the next day. The trip home is uneventful; she feels bad, but she makes it. Since it is summertime, she can rest and not worry about school.

Unexpectedly, the very next week, another pain episode strikes. Again, there are no identifiable triggers. The pain is severe, and after no relief from her home regimen, she uncharacteristically asks to go to the hospital. Discharged two days later, she is now on NSAIDS, an escalated opioid regimen and using crutches to keep weight off the left knee due to severe pain. Her doctors cannot explain why her disease is suddenly so active.

***Strength.*** A trip is already planned for a family wedding in Houston the day after her discharge and she is set on going. She is looking forward to seeing her sister and cousins; they already had several fun events planned. Using a wheelchair and crutches, she makes the flight, and over the next 5 days, she takes numerous doses of alternating pain medications. She attends all events—she smiles, she laughs, sometimes she winces and sometimes she cries. She is frustrated, but hides it well. She tries to find a comfortable position; there is not any. The morphine causes urinary retention—a new problem—she deals with it. She is not comfortable but she is happy. Instead of hearing later about all the fun she missed, she is part of it; she is with her family and living in the moment.

***Resilience.*** This is a story of a teenage girl determined to live a normal life in spite of any odds life may have dealt her. She does not want to lie in bed for days, wallowing in self-pity while waiting for the pain to pass. Even when in pain, she wants to be able to be as active as she can. Like most teenage girls, she wants to go to college, get a job, get married, have kids, and grow old. She wants to be normal.

***Reality.*** This teenage girl is my daughter. As a general internist with a practice that includes adult sickle cell care, I worry about her future. What happens when she becomes an adult? Will she become a statistic; stereotyped, stigmatized, and judged because she may rely on narcotic pain medications to live a normal life? Despite the challenges of this disease, I have learned so much, making me a better physician and hopefully a better person. In her early years, I would avoid giving her narcotics. I remember not wanting to take the first morphine prescription written for her; I did not want my child to be someone on narcotics. Whenever she was in pain, I would give her ibuprofen, wait for hours before adding hydrocodone/acetaminophen and finally, reluctantly, morphine if the pain persisted.

One day, it dawned on me: why was I the one deciding what type of pain medication she needed? I was imposing my stereotype about narcotic pain medications on my child. Instead, I should teach her to advocate for herself. From that point forward, when in pain, I ask her what pain medication she needs. She uses the 10-point pain scale to aid in her decision-making. Most of the time, it is Ibuprofen, sometimes hydrocodone/acetaminophen, and rarely she will ask for morphine right from the start because the pain is severe. I encourage her to speak directly to her doctors even when she is sick and ask them questions. We have candid discussions about narcotic medications, the risks, potential for abuse, and her responsibility to keep them safe.

***Hope.*** I have taught my daughter to own her disease; she is learning to not let it control her life. She has taught me to never judge my patients, to trust them first and care for them always. She leaves for college soon. It is difficult to realize that I will not always be there when she gets sick, to comfort her and tell her everything will be okay. This has been a life-changing and life-enriching journey for both of us—and will continue to be.

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***Compliance with Ethical Standards:***

***Conflict of Interest:*** *None.*