Transitions of Care in Chronic Disease

A Patient with Sickle Cell Disease

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Learning Objectives

- Identify protective and negative social determinants of health for patients with chronic disease during and after transition of care from pediatric to adult care.
- Explore the multiple members of a sickle cell patient's community who contribute towards his/her health successes.
- Describe the impact of unconscious biases in the patient-provider relationship for patients with sickle cell disease.
- Explore possible collaborations between Primary Care Physicians (PCPs) and specialists to better coordinate care for patients with chronic disease.

Case synopsis

William is a 22 y/o male with sickle cell disease (HbSS) complicated by two prior episodes of acute chest syndrome and cholecystitis status post cholecystectomy, now presenting with right sided chest pain concerning for recurrent acute chest syndrome. Upon admission, he was afebrile, tachypneic, and hypoxic with oxygen saturation 88% at room air. Notable labs included a leukocytosis, elevated LDH, and elevated reticulocyte count. A chest x-ray showed a right lower lobe infiltrate. He was started on antibiotics and a patient-controlled analgesia (PCA) pump and transferred to the ICU for exchange transfusion. He was subsequently transferred to the general medicine floor where he was weaned from the PCA pump and discharged after one week.

William has had a sickle cell crisis once every few months which has led to eight hospitalizations over the past three years, of which two were for acute chest syndrome. He followed with a hematology physician but has missed several appointments over the past few months after moving in with his parents who live an hour and a half away from the city. He used to have a PCP but has not seen her in over five years because he felt like he had a better relationship with his hematologist, and it was easier to just have one provider. He is not interested in taking hydroxyurea because he witnessed his older sister, who also has sickle cell disease, have increase nausea and abdominal pain while on this medication and he did not feel like it reduced the frequency of her sickle cell crises. He is frequently frustrated when providers push him to start this medication and endorses hesitation in coming into the hospital due to providers judging the way he chooses to live with his disease. He describes the emotional struggles he goes through with each hospitalization and feeling embarrassed and nervous to talk about his health experiences with his friends. He is also hesitant to transition to the adult hospital after witnessing negative experiences his sister has had with communicating her pain and medication dosing to emergency room providers.

William graduated from an art institute and is currently a graphic designer making flyers and invitations. He is self-employed so it has been difficult to find consistent work. He recently moved in with his parents due to financial difficulty and has trouble commuting into the city where his physicians are located. He endorses marijuana and social alcohol use. He reports having a good support system at home.

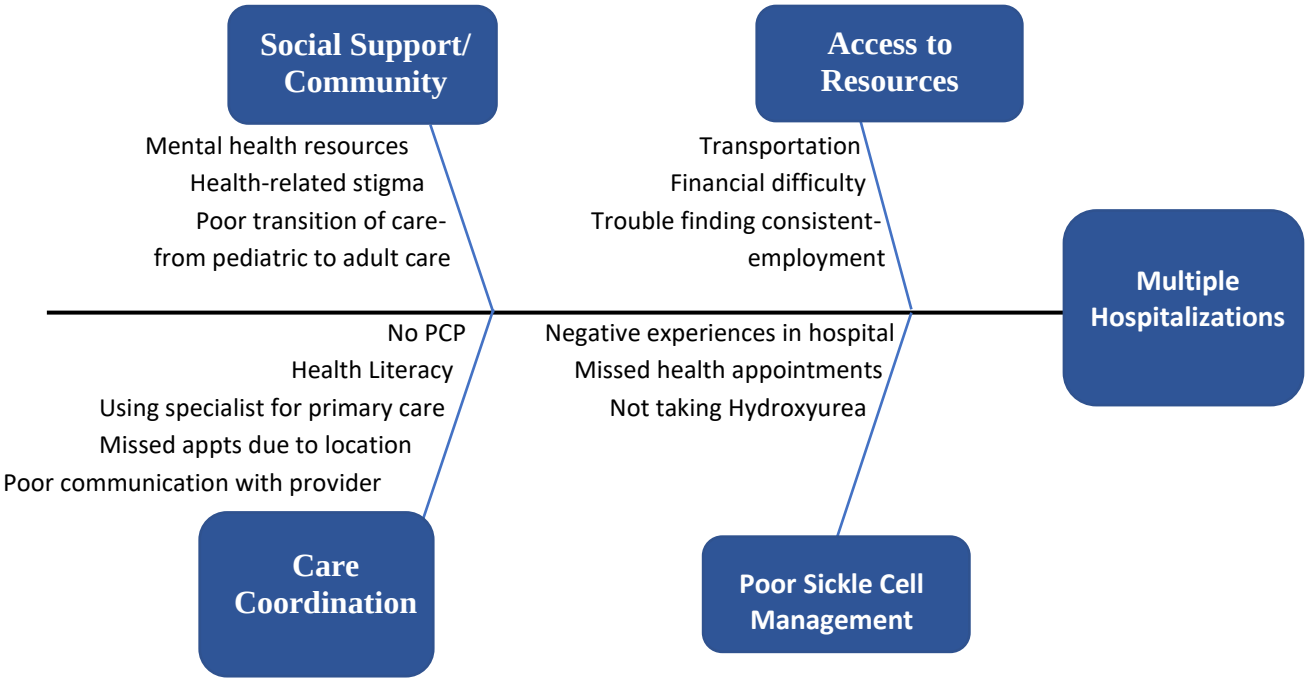
Questions:

1. **Construct a problem list for this patient**
2. **Conduct a root-cause analysis for at least one problem**
3. **Describe the positive/protective social determinants of health for this patient**
4. **Describe negative social determinants of health factors**
5. **Propose patient-level solutions with attention to facilitators and barriers**
6. **Imagine possible health system or institutional solutions**
7. **Discuss potential community/societal-level solutions**

Facilitator's Guide:

1. **Construct a problem list for this patient**
    a. Acute Chest Syndrome
    b. Recurrent Sickle Cell Crises
    c. Multiple Hospitalizations

2. **Conduct a root-cause analysis for at least one problem**



Several factors contribute towards William's recurrent hospitalizations. His recent move into his parent's home has led to difficulty with transportation into the city for appointments. The increased distance from the hospital has made it hard to attend all of his health care appointments and therefore has not seen his PCP in several years. His difficulty in finding a steady job has also led to financial dependence on his parents. He is fearful of the adult hospital system because of his sister's negative experiences and feeling like he is not prepared to be completely independent. He struggles with the lack of peer support and patient advocacy from health professionals and feels like he is judged for the decisions he makes about his healthcare. A combination of these factors contributes to his increased disease severity and multiple hospitalizations.

3. **Positive/protective social determinants of health for this patient**

William has a very strong social support system through his parents and siblings. They are able to provide not only emotional but also financial support throughout this process. He also has health

insurance and has been able to pay for his medical care. He has access to primary and specialty care. He has a degree. He has access to safe housing and food.

4. Describe negative social determinants of health factors

William is currently self-employed, so he does not have a stable source of income. Although he has family to drive him sometimes, they are not always available. He does not have a license or car so he cannot drive himself. He also does not always have enough money to pay for a bus ticket or taxi. He is living with a chronic disease.

5. Propose patient-level solution with attention to facilitators and barriers

The patient level solutions that he could take include setting up a schedule ahead of time with family and friends who are able to provide transportation during his PCP and specialist visits. This may increase the chances that he does not miss an appointment due to lack of transportation. Often, insurance programs, especially Medicaid, offer transportation services that allow some patients to get to their medical appoints. Determining if this patient is eligible for this type of assistance would be beneficial. If transportation is still a barrier, his care team can help him find a PCP closer to him and eliminate distance as a barrier to care. Alternatively, attempting to coordinate his PCP and specialist visit on the same day would also increase the chances of making both appointments.

Improving communication between patient and provider is key in strengthening the relationship, engendering trust and improving patient outcomes. This can be done by encouraging a shared decision-making model in which physicians and patients spend an equal amount of time talking and increase the patient's involvement in his/her own health care decisions[1]. Higher health literacy is associated with more positive health behaviors and improved clinical outcomes in adults[2]. Taking the time to better understand the health literacy level for each unique individual will allow for more personalized education. In order to facilitate this model, the physician should work to recognize their patient's perception and understanding of his/her disease as well as the individual as a person. In William's case, his strength is art so asking him to share a piece of art that depicts his journey with sickle cell disease could offer valuable insight into the unique obstacles he faces.

Lastly, he is passionate about graphic design and his artwork so he may want to continue searching for full time or part time jobs locally that can help him gain some financial independence. Having familial support is a major facilitator towards helping him get back on his feet.

6. Imagine possible health system or institutional solutions

Health system solutions could include providing some means of transportation support (i.e. bus passes) to help with transportation to and from appointments. Difficulty with transportation is a common reason why patients are unable to make their appointments with their PCP or specialist so finding ways to combat this barrier would be incredibly helpful for many patients.

Improving the transition process from childhood to adulthood would also provide significant benefits. Young adulthood is a high-risk time for increased morbidity and mortality largely due to poor transition of care from a patient's pediatrician to adult hematologist/oncologist[3]. This leads to lack of proper follow up with their PCP and specialist, decreased use of hydroxyurea, and/or increased hospitalizations[4]. Creating an improved transition program that empowers young adults to take control of their health is important. A consensus statement by AAP, AAFP, and ACP articulated 6 critical first steps for successful transition which includes (1) having an identified health care professional that will help with care coordination and current/future health care, (2) identifying the skills and knowledge needed to provide developmentally appropriate healthcare transition services to adolescents with chronic disease, (3) maintain an up to date medical summary that is easily accessible, (4) developing an up to date transition plan that is done in collaboration with patient and their families, (5) ensure standards for primary and preventive health care are applied to adolescents, and (6) ensure affordable and comprehensive health insurance is available as patients transition from pediatric to adult hood[10].

Combating unconscious biases within the health system is important for all patients but specifically the sickle cell population. Focus groups with young adult and adult sickle cell patients have shown that negative biases experienced in the emergency department or hospital are some of the reasons why they dislike the healthcare system[5]. Patients have described situations where providers hesitate to give them pain medication, extended waiting times in the emergency department, and general discomfort seeking out care independently[5]. Increasing unconscious bias training and how language can impact a patient's experience within the hospital system is an important measure that hospitals can take.

Lastly, offering telemedicine services to individuals in rural communities has potential to increase access and continuity of care[11]. Some of the benefits of telemedicine include increased frequency of encounters with specialists, continuity of care, decreased travel and waiting time[11]. In addition, this patient might also benefit from a provider home visit. A home visit not only would provide insight into this patient's home environment and neighborhood, but also allow for provision of medical care and review of medications.

7. Discuss potential community/societal-level solutions

Some community level solutions include creating a support system and fostering an environment in which discussing sickle cell disease (SCD) is not ostracizing. Organizations like the Children's Sickle Cell Foundation in Pittsburgh has been powerful in supporting families who have children with sickle cell disease. This program provides emotional support, educational help, and an overall community that supports each other through this process. Peer support groups can promote peer encouragement, confidence building, and exchange of experiences and information.

A common barrier identified for transition programs is the ability to identify an adult provider that specializes in sickle cell disease[12]. Due to the lack of specialists, many patients are then transferred to a general internist who are not always comfortable in taking care of patients with sickle cell

disease. This is a barrier to creating a medical home for young adults that are recently transitioned out of pediatric care. Focusing transition research on ways to promote and increase education and expertise on sickle cell care would be an important next step.

Implementation of community health workers (CHWs) could be a valuable asset to helping sickle cell patients navigate the health system and provide social support. Specifically, CHWs are non-clinicians who work with underserved, medically complex individuals to improve patient health outcomes[13]. CHWs are impactful on various levels of the socioecological model starting from medication adherence and individual coping to improving economic self-sufficiency to better understanding reproductive choices to navigating multiple health appointments and transportation. CHWs can also help with navigating the work and school system for missed days due to illness. Case managers or service coordinators can also provide these services.

Lastly, funding for sickle cell research is significantly reduced compared to other chronic diseases. For example, a 2013 study published in the journal Blood found that funding per individual was about 7-11 times greater in 2011 for cystic fibrosis than SCD and 3.5-fold higher NIH funding. Moreover, 5 new drugs were approved between 2010-2013 for cystic fibrosis compared to none for SCD[14]. Increasing advocacy for more funding and research is an important long-term step to not only finding more therapies but also in improving health outcomes for SCD patients.

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