Management of Sickle Cell Disease: Consolidated CER Themes and Questions

Questions for Care Transitions breakout

Care delivery models

Personnel-focused

- What are the comparative benefits and risks of a certified peer support specialist, social worker, and nurse navigator on the transition from childhood to adult care for sickle cell disease?
- Can community health workers improve continuity and care transitions for sickle cell, especially patients heavily affected by social determinants of health?
- What is the comparative effectiveness of a structured transition program, initiated by age 13, with the adult provider team only, community-based organization only, or both, on a "successful transition" from pediatric- to adult-focused care in adolescents and young adults with sickle cell disease.
- Does age of transition (18 vs 21 years) of sickle cell patients to adult care lead to better outcomes as measured by decreased ED visits and hospitalizations?

Care-setting focused

- Is there decreased ED utilization or increased patient-reported quality of life among young adults seen in a comprehensive SCD center with lifelong care compared to the more traditional, separated, pediatric and adult hematology care model?
- What are the comparative benefits and risks of a sickle cell ACA Medicaid Health Home model of care and a hospital-based sickle cell clinic in transitioning from adolescent to adult care for people with sickle cell disease?
- Will transition programming that focuses on establishing a medical home (primary care driven) be more effective in reducing acute care utilization and access to disease modifying therapy than programming focused on specialty hematology care?
- Upon discharge home from the emergency department, what is the comparative effectiveness of different referral strategies to improve quality of life indicators (ASCQ_Me measures), in persons with SCD?

Clinician training

- What is the comparative effectiveness of different educational mechanisms such as guidelines, multimedia presentations or publications that can be utilized to train primary care providers in pain management of sickle cell patients?

- Will establishing regional clusters of medical neighborhoods for SCD adults that link a PCP with a hematologist with additional practice support versus traditional provider CME/written education material versus virtual peer mentoring via "Extension for Community Outcomes ECHO" for both primary care providers and adult hematologists be more effective in increasing access to adults with SCD and yield reduced acute care utilization and increased access to disease modifying therapies?
- What is the effectiveness of a standardized questionnaire or guideline to address transition from pediatric care to adult care?

Self-care management

Peer navigator vs. technology-based interventions

- What is the comparative effectiveness of patient navigation versus mobile health coaching app interventions on successful engagement in care for individuals with sickle cell disease transitioning from pediatric to adult care?
- Among young adults with sickle cell disease who will transition from pediatric to adult care, would peer navigators compared to text messaging/apps improve disease self-management (e.g. knowledge of disease, health related quality of life) and improve health promoting behaviors (e.g. attendance at outpatient visits, communication with providers, and medication adherence)?
- What is the comparative effectiveness of video games versus one-on-one health coaching on successful engagement in care for individuals with sickle cell disease transitioning from pediatric to adult care?
- What are the comparative benefits and risks of patient navigators, technology-enhanced patient-centered programs, and patient-provider learning networks on self-management in children with sickle cell disease preparing to transition from pediatric to adult care?

Patient activation interventions

- Compare patient activation and engagement models on improving transition of care for patients living with sickle cell disease.
- Among young adults with sickle cell disease who will transition from pediatric to adult care, would group visits compared to individualized patient visits: 1) improve knowledge about hydroxyurea and management of acute symptoms of their disease; 2) improve medication adherence; and 3) decrease emergency department utilization?
- What are the comparative benefits and risks for care continuity from utilizing electronic self-recorded pain diary records as a communication aid with the adolescent SCD population transitioning to adult care?
- Among adolescents with sickle cell disease, would a disease-specific transition readiness curriculum (e.g., Sickle Cell Disease Treatment Demonstration Program Transition Curriculum) compared to a general transition readiness curriculum (e.g., Got Transition) enhance self-efficacy to manage symptoms of their disease and improve health promoting behaviors (e.g. attendance at outpatient visits, communication with providers, and medication adherence)

Questions for Pain Management breakout

Self-care management

- In adolescents and adults with sickle cell disease, do individualized pain plans for acute pain crisis jointly developed by patient and provider compared to standard order sets/algorithms implemented in the emergency department, lead to more timely administration of pain medication, improvement in pain (e.g. decrease in pain score), and decreased hospital admissions due to acute pain crisis.
- In people with sickle cell disease, do individualized home pain plans compared to cognitive behavior therapy including mindfulness interventions lead to improved coping, decreased missed days from school or work and decreased emergency department visits for acute pain crisis?
- What are the impacts on pain control and vaso-occlusive crises resolution from utilization of smart phone (tracking and communication) application(s) for children experiencing painful episodes with SCD?

Pharmacologic pain management

- What are the comparative benefits and risks of opioids and non-opioids in the treatment of pain in patients with sickle cell disease? Chronic use of short-acting opioids and slow-release opioids? What quality of care and drug safety parameters should be assessed?
- What are the comparative benefits and risks of transdermal buprenorphine (an opioid partial agonist) and full opioid agonists in the treatment of individuals with acute painful sickle cell crisis?
- What are the comparative benefits and risks of hydroxyurea therapy vs. alternative drug therapies on pain, quality of life, physical functioning, and clinical outcomes for patients with sickle cell disease?
- What is the comparative effectiveness of short door to initiation of pain medication times in all adults with sickle cell disease in academic and community hospital settings?
- Is intravenous fluid superior than oral fluid for improving patient outcomes (decreased pain, decreased need for opioids, decreased need for admission) for patients with sickle cell disease in an acute pain crisis?
- What are the comparative benefits and risks of shared-decision making, personalized patient decision analysis, and patient education on hydroxyurea use in patients with sickle cell disease?

Pharmacologic vs. non-pharmacologic therapies

- What is the comparative effectiveness of a variety of pharmacologic and non-pharmacologic interventions for the treatment of vaso-occlusive crisis in the emergency department setting for patients with SCD?
- Does the implementation of non-pharmacologic measures such as Art Therapy for patients with Sickle Cell Disease admitted with acute VOC increase satisfaction with pain management and time to acceptable analgesia as measured by pain scales, and decrease the quantity of opioids

used throughout the hospitalization as well as length of stay compared to management with opioid therapy alone?
- What is the comparative effectiveness of cognitive behavioral therapy and non-opioid pain management versus chronic opioid therapy on pain experience for adults with sickle cell disease?
- Do acute pain management protocols including non-pharmacologic techniques such as heat, water therapy, acupressure and biofeedback result in improved patient satisfaction or lower opioid analgesic equivalent usage when compared to protocols that do not include non-pharmacologic approaches to pain control?

Improving health care processes

Among care settings

- What are the comparative risks and benefits of pain treatment in emergency departments, inpatient observation units, day hospital facilities (e.g. Sickle Cell Day Unit) in the management of acute pain in adults and children with SCD?
- Can individualized patient care plans accessible to both primary providers and emergency providers improve the management of patients with sickle cell disease, including decreased emergency department utilization and decreased episodes of acute pain crisis?
- What are the comparative benefits and risks of virtual human technologies provider training, face-to-face provider training, and telementoring provider training on the treatment and management of acute pain crises in patients with sickle cell disease?

Within care settings

- What are the comparative benefits and risks of implementing health systems-level initiatives in the emergency department on timeliness of pain management for patients with sickle cell disease?
- What is the best way to manage pain in an ED that does not know the patient?
- What are the comparative risks and benefits of global functional pain management models as an alternative to the classic "chronic/ acute pain" models for adults and children with sickle cell disease?