The Emotional Impact of Sickle Cell Disease

Temeia Martin, MD
Medical University of South Carolina
Departments of Internal Medicine and Psychiatry
April 22nd, 2017

Disclosures:
Dr. Martin has documented that she has no disclosures.
Objectives:

- To explore emotional aspects of sickle cell disease.
- To understand how depression, anxiety, and cognitive disorders affect patients with sickle cell disorders.
- To overview strategies and interventions to address emotional and cognitive symptoms of patients with sickle cell disorders.

Introduction:

- Cognitive disorders
- Mood disorders (adjustment disorders, depression)
- Anxiety
- Substance abuse
- Thought disorders (psychosis, schizophrenia)
Introduction:

The occurrence of mental health disorders is influenced by:
- living with a chronic stigmatizing disease associated with chronic pain
- unpredictable painful crises
- multiple serious complications
- poor health-related quality of life (HRQOL)
- problems of pain management
  - Under-treatment
  - Potential for substance abuse
- coping styles
- central nervous system injury
- high mortality

Dr. Levenson is professor in the Departments of Psychiatry, Medicine, and Surgery, chair of the Division of Consultation-Liaison Psychiatry, and vice chair for clinical affairs in the Department of Psychiatry at Virginia Commonwealth University School of Medicine in Richmond.

Introduction:

- Health related quality of life (HRQOL) – an individual's or a group's perceived physical and mental health over time.
  - Similar to dialysis population
  - Significantly worse than general population
  - Worse than cystic fibrosis
Introduction:

Why is this important?

◦ Self-management
◦ Disease outcomes (treatment adherence)
◦ Utilization of acute care services/length of stay (LOS)
◦ Quality of life
◦ Social, economic, and healthcare disparities

Challenges:

› Lack of data and research
› Society's attitudes to SCD and those affected
› Limited evidence based interventions and guidelines
› Variability in coping styles
› Maintaining adequate relationships with healthcare providers
Introduction and background:

- The NHBLI released a comprehensive summary of recommendations for SCD management in 2014.
- This summary did not address specific information pertaining to diagnosis and treatment of psychiatric co-morbidities in SCD.

Depression and SCD:

- Rates similar to those found in other serious chronic medical disorders
- 18% to 44%, varies amongst studies
- Influenced by:
  - chronicity of the illness
  - unpredictability of crises
  - chronic pain
  - overwhelming nature of medical complications
  - substantially reduced life expectancy
  - racial prejudice and stereotyping
  - chronically prescribed opioids
Opioids and depression:

- Opioids are central nervous system depressants and chronic use may induce depression.
- An association between mental illness (substance use, anxiety, and depression) and opioid misuse exists.
- Diagnoses related to opioid induced mood disorders are categorized in the DSM-V.

Depression and SCD:

- SIGECAPS

  - FATIGUE, IRRITABILITY, NEGATIVE THOUGHTS, and INSOMNIA

- Episodes of depression in childhood and adolescent years increases the risk of recurrence in adults with SCD.

- Rates of suicide attempts and suicide completion are no greater than that seen in the general population.
Depression and SCD:

- PiSCES Project:
  - 27.6% of adults with SCD were depressed
  - pain on significantly more days than non–depressed subjects (mean pain days=71.1% versus 49.6%, \( P<.001 \))
  - On non–crisis days, depressed subjects had higher mean pain, distress from pain, and interference from pain than those without depression
  - Depressed subjects had poorer functioning on all dimensions of HRQOL

- Research to date has shown variable findings.
  - Systematic literature review conducted by Charles R. Jonassaint et al
    - 12 studies; 7 studies found a significant association between depression and utilization, 5 did not
    - Depressed patients had an estimated 2.8 times greater relative risk of higher utilization compared to patients without depression
    - Depressive symptoms are common and may increase risk for poor outcomes and utilization
    - Small sample sizes, retro–prospective designs, and short intervals of follow–up
Anxiety is associated with more pain
Anxiety is correlated with VOC
Anxiety is associated with poorer health-related quality of life
No clear relation to increases in urgent health care utilization
Cognitive Disorders are defined as a category of mental health disorders that primarily affect learning, memory, perception, and problem solving.
Cognitive Disorders in SCD:

- "Neurocognitive dysfunction may be the most important and least studied problem affecting this aging population. To our knowledge, controlled studies of neurocognitive function in adult patients have not been reported and routine screening after childhood is not performed."
  - Elliott P. Vichinsky, MD, of Children's Hospital & Research Center in Oakland, California.
  - 12 sites, 149 subjects ages 19–55 and 48 healthy adults
  - Older age, lower Hgb
  - Imaging findings do not explain the difference

- More common in adults with minimal complications: intellectual ability, attention, short-term memory, processing speed (National Heart, Blood, and Lung Institute, JAMA, 2010)

Cognitive Disorders in SCD:

- Strokes
  - 17 percent of SCD children under the age of 14 have silent strokes and the rate increases to 23 percent by the age of 18, with the size and number of lesions increasing.

- Silent brain injury
  - Neonatal hypoxia
  - Cerebral ischemia from vasoocclusion
  - Stenosis of cerebral arteries in asymptomatic children and adults

- Delayed mental development
- Seizures
Cognitive Disorders and SCD:

- Silent brain injury
  - Neonatal hypoxia
  - Cerebral ischemia from vasoocclusion
  - Stenosis of cerebral arteries in asymptomatic children and adults
  - Chronic hypoxic damage to the brain or diminished pulmonary function
  - Sub-acute brain damage that occurred during bouts of hypoxia associated with events such as aplastic crisis, acute chest syndrome, and obstructive sleep apnea
  - Chronic nutritional deficiency associated with increased metabolic demands.

Cognitive Disorders in SCD:

- Study of 393 patients Sickle-Cell Disease Contributes to Cognitive Impairment in Children
  - Acacia C. Grimes university of Missouri
  - 255 with hemoglobin SS and 118 with hemoglobin SC
  - Ages 4–16
  - Wechsler Intelligence Scale for Children (WISC–R or WISC–III) and the Woodcock–Johnson Math and Reading Achievement Tests, Full–scale IQ (FSIQ) and an MRI
  - Children and adolescents with SCD between 6 and 18 years of age have a significant deficit in neuropsychometric performance associated with silent infarction and a significant decline in performance with age in certain areas
  - Children with sickle cell disease coupled with silent infarcts do present cognitive complications in their complete cognitive capacity specifically in verbal comprehension, perceptual reasoning and working memory
Cognitive Disorders in SCD:

- Ongoing research to determine if transfusions, neuroprotective agents, Hydroxyurea, and/or oxygen may offset these findings
- Identifying those at risk and the role of cognitive rehabilitation programs
- Standardized and comprehensive neuropsychological assessment

Conclusions:

- Literature findings are inconsistent.
- Little is known about the actual impact of mental health factors on disease outcomes or the impact of SCD on mental health.
- Diagnosis and treatment of mental illness is necessary to improve outcomes in patients with SCD.
- Medical advances that address anemia, infection, and sickle cell crises have not significantly decreased the risks for development of depression and anxiety.
References: