Supporters of Families with Sickle Cell Disease, Inc.

Oklahoma Sickle Cell Community
Embracing Change Together
October 2020

Pat L. Corley, RN -Sickle Cell Nurse Educator

Velvet Brown Watts, MSW, CM-Social Worker

Shaquanda Lewis, Patient Advocate

Lindsey Corbitt, Community Health Worker

The Mission of Supporters of Families with Sickle Cell Disease is to increase self-efficacy and improve the overall quality of life for individuals living with sickle cell and its inherited disorders and traits, impacting children, adults and their families within Oklahoma, through systemic changes in patient care, disability policies, education, family support, economic self-sufficiency, awareness and advocacy.

Our services are provided without discrimination by age, race, ethnicity or gender, and we serve everyone without any discrimination, including individuals who are part of the LGBTQ, at-risk youth, disabled and veteran communities.

Supporters' is the community-based contractor for Oklahoma Health Care Authority and the State Lead for the HRSA Sickle Cell Disease Newborn Screening Program.

Objectives

At the end of this session participants will have a better understanding of:

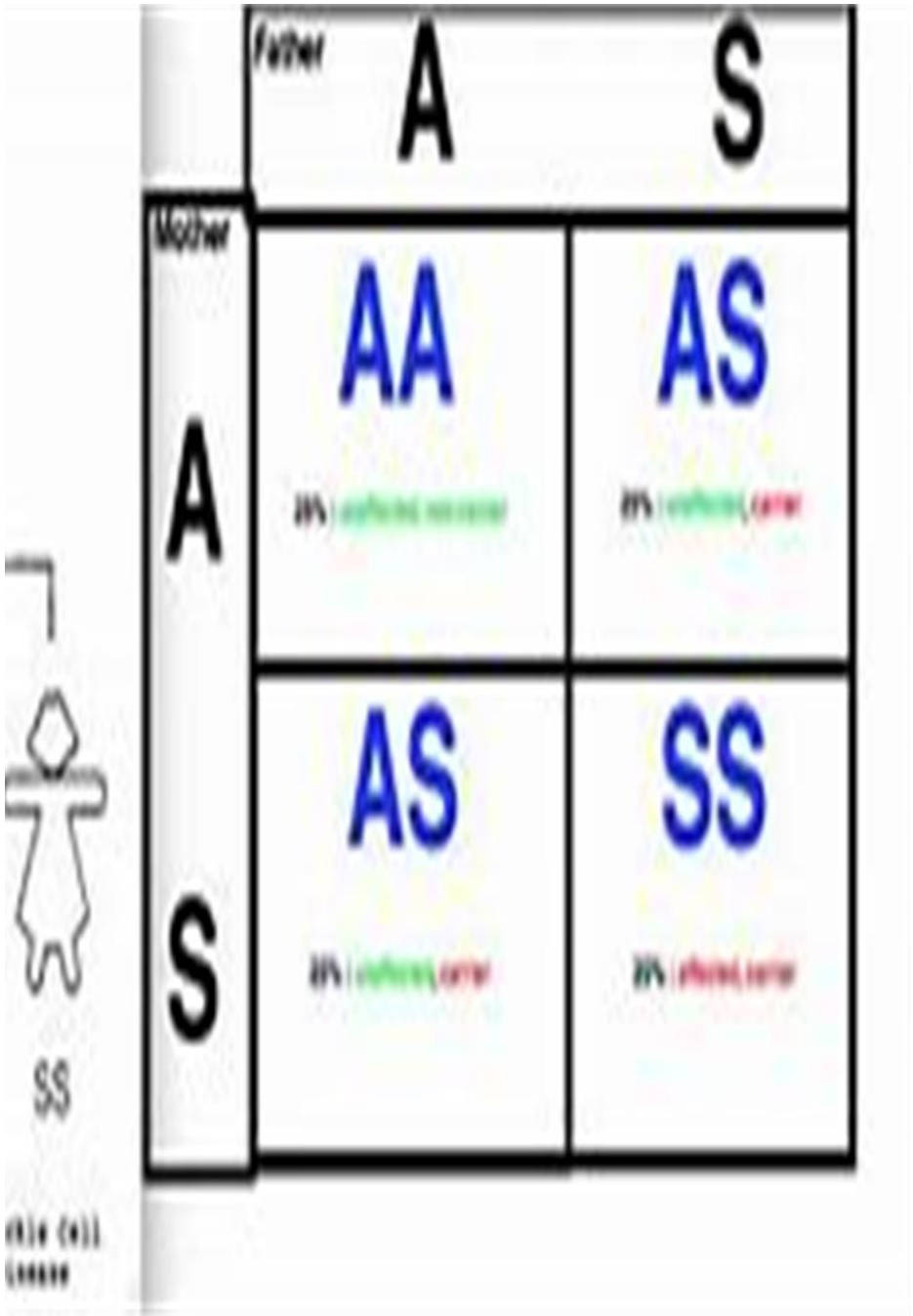
- **Sickle Cell Disease and Sickle Cell Trait**
- **Impact of disease**
- **The community's experiences, needs, concerns, challenges, and barriers related to access to care**
- **New treatments/ medicines**
- **NIH SCD Best Practices / Guidelines**
- **Ways to engage families in the journey in receiving optimal care and improving health outcomes**

Medical Trauma

Pediatric medical traumatic stress refers to a set of physiological responses of children and their families to serious illness, medical procedures, and invasive or treatment experiences. These responses may include arousal, re-experiencing, and / or avoidance.

National Child Traumatic Stress Network

www.nctsn.org/trauma-types/medical-trauma



Sickle Cell Trait

- Carrier State of the disease
- Single copy of S gene +(normal A Gene
- 3 Million Americans diagnosed with Sickle Cell Trait
- 10 % African – American population
- 1 in 13 African – American born with Sickle Cell Trait (hemoglobin AS)

Sickle Cell Disease

- group of red blood cell disorders
- sickle gene+ abnormal hemoglobin gene
- Inherited
- Autosomal recessive
- Beta chain substitution
- 6th position of hemoglobin molecule
- valine substituted for glutamic acid

Impact of SCD

Genetic (inherited)

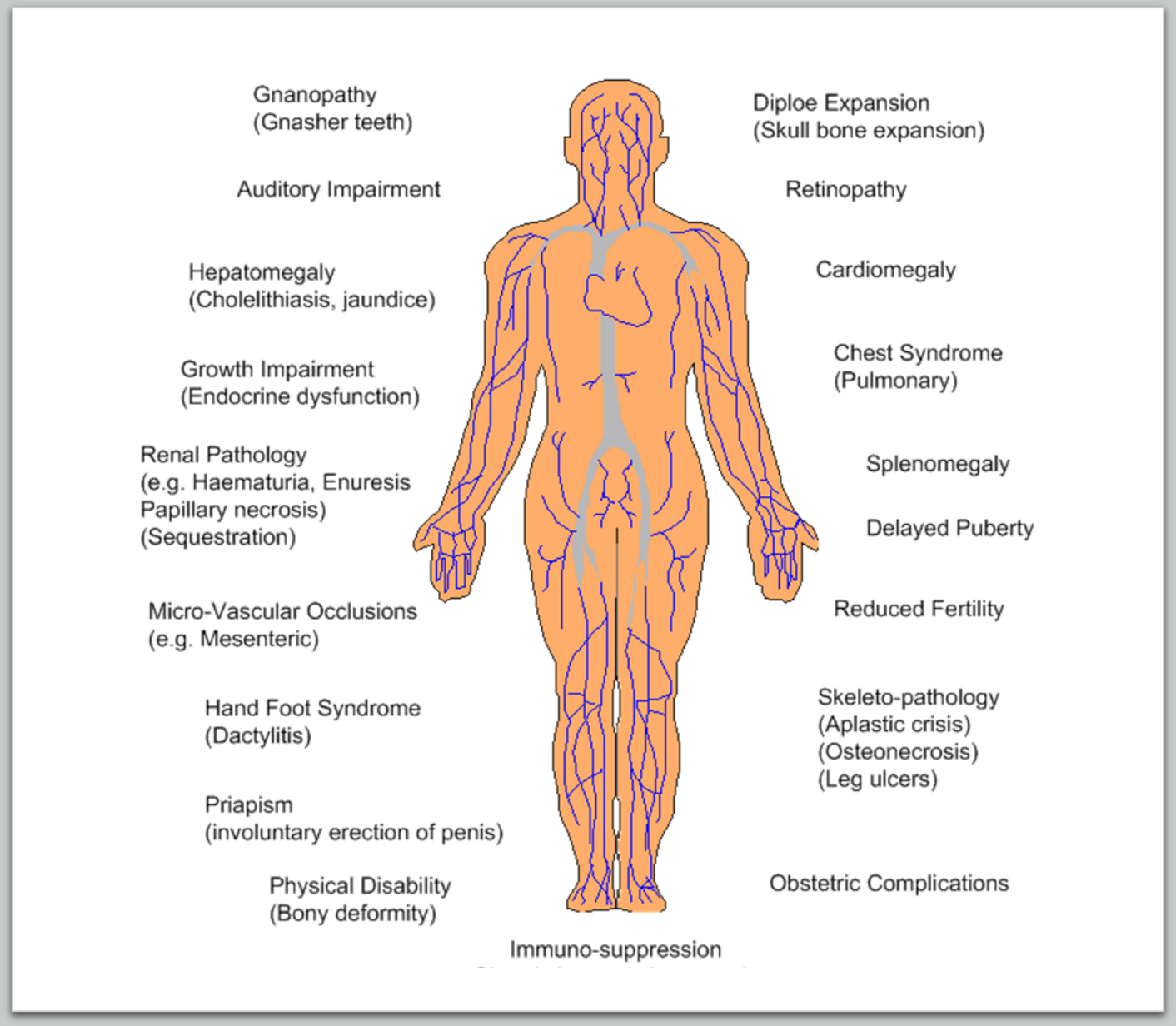
Chronic (lifelong)

Unpredictable

Potential for serious complication

Public health problem

Clinical Complications of Sickle Cell Disease



What is the Earmark of SCD?

Pain

- from clumping of RBCs
- sticking of RBC to lining of blood vessels

“Pain is the leading reason people with SCD most often come to the hospital”

Patient Challenges

- **Trauma due to past and present experiences**
- **Judgements and stigmas**
- **Devaluing / Diminishing pain and journey experiences report**
- **Providers and care teams that lack knowledge and understanding about SCD, care and pain management, and national best practices**
- **Poor communication skills**

Provider Comments

- **“ I don't really believe they're in pain, they're here again, they're just drug-seeking.**
- **” Hearing those kinds of things really makes them feel badly.**
- **“He's heard those kinds of things when he's come in. (Provider for EHHU-4)**
- **We have ended up creating a whole cadre of *addicts*, and their lives appear to me**
- **“to be one constant quest of narcotics. (Provider for EHHU-1)**
- **I don't know if there is a chronic sickle pain crisis or if it is all addiction.**
- **I know that I don't know that. (Provider for EHHU-**
- **This is a chronic pain syndrome**
- **You are going to have to deal with it.**
- **Dealing with it means learning relaxation techniques,**
- **learning how to use pain medications wisely …**
- **Unfortunately, the attitude that one is going to**
- **zero pain a hundred percent of the time, basically means**
- **you're going to be a dope addict. (Provider for EHHU-7)**
- **The more often they come in, the more [patients] get stigmatized**
- **and the more they get labeled as “drug-seeking,” “they're here again”**
- **and [staff] don't care whether the patient hears. Especially in the emergency**
- **room, some of the nurses and doctors talk about them within their hearing: “**
- **Why won’t they listen to me**

Sickle Cell Pain is Real!

SCD pain can cause patients to experience:

- Fear of Death
- Effects
- Central sensitization
- Hyperalgesia
- Altered opioid metabolism
- Chronic pain
- Mood changes
- Emotional disturbance
- Behavioral dysfunction affects of the patient's life
- A crisis in the life of family members friends, and colleagues

What is Sickle Cell really like?

Painful

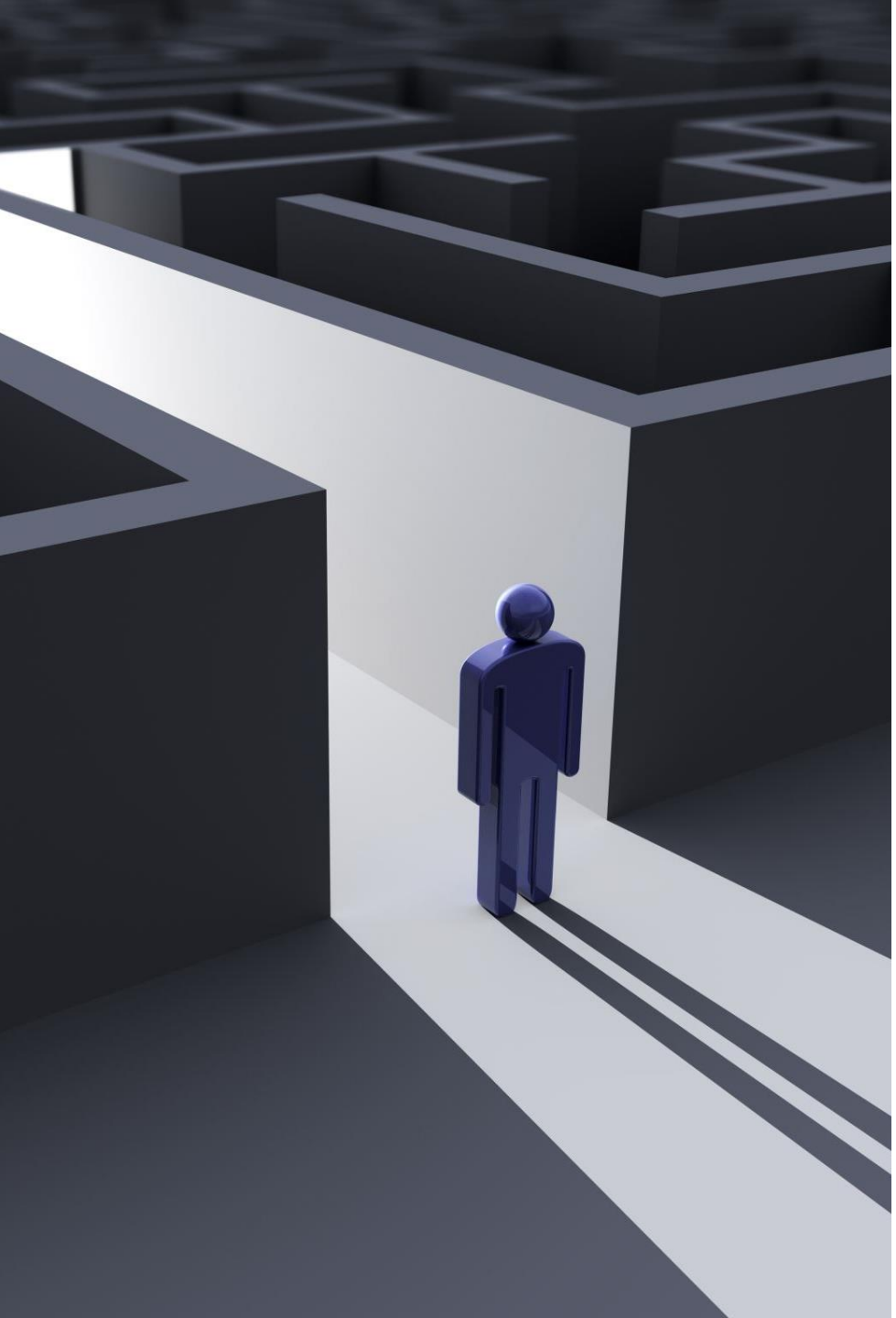
- The most common manifestation of SCD is vaso-occlusive crisis
- Crisis characterized by intermittent, unexpected episodes of excruciating pain.
- Crisis episodes often come on suddenly, SCD care is influenced by many factors.
- Pain is intense
- Highly Variable
- Patients have their own personal pain pattern
- Elusive in nature
- Usually not life threatening
- People with SCD can experience acute, chronic and neuropathic pain at same time
- Less than 1% addiction rate
- Most patients manage their pain at home

Forms of Barriers to Treatment

- patient behavior resulting from trauma of past experiences
- opioid epidemic (67.57%)
- overcrowding (64.86%)
- concern about addiction (49.55%)
- lack of SCD care protocols
- poor communication
- stigma and judgement
- provider knowledge
- provider bias.

- Belief related to manifestation of vaso-occlusive pain
- Sociocultural factors affecting pain assessment
- concerns regarding addiction and pseudo-addiction.
- Minority community
- Community of color/BIPOC
- Underserved community Misunderstood and devalued
- Historical disrespected and disregarded.
- Underprivileged
- Lack of cultural intersectionality

Addressing these barriers will ensure that people with SCD have their pain managed more effectively, improve their quality of life and potentially reduce A&E attendances and admissions to hospital.

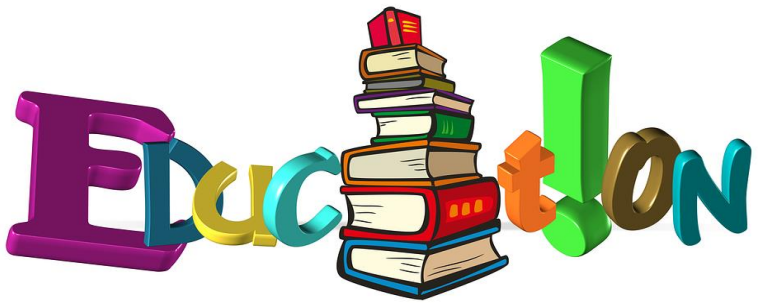


Psychosocial Barriers to Successful Management

- Educational
- Social
- Mental Health/Emotional
- Occupational/Employment
- Financial/
- Spiritual
- Housing
- Several aspects of the social determinants of health

Question: Why do you think there are such barriers in these areas and how can you and your practice mitigate these?

Children with Sickle Cell Disease miss an average of 29-31 days of school per year



- Specialists appt.
- Chronic Transfusion appt.

Acute Episodes of Illness

- Hospitalization

Home

- Recovering from recent hospitalization
- Too sick to attend school, but not sick enough to require hospitalization
- 37% of children with Hb SS have had silent strokes on MRI by age 14 years .
- Learning problems and mental health issues may not be prioritized during routine visits for children with SCD
- SCD complications may be prioritized
- Lack of knowledge about silent strokes (providers, parents, teachers)
- Parents do not recognize problems

Teaching Points

Sickle Cell Pain

Understand	Understand potential VOE triggers and how to avoid them
Assist	Assist patients who better understand chronic pain, mixed pain presentation, unrelated persistent pain
Ensure	Ensure patients understand how to take the pain medicines prescribed to manage their pain.
Help	Help patients understand the importance of hydration in preventing VOEs , increasing plasma volume and reducing blood viscosity
Instruct	Instruct patients on use the incentive spirometer every hour to prevent blood clots and pulmonary complications.

Nurse's comprehensive pain assessment and education

Pain……..

- location
- intensity
- duration of the pain episode
- what normally relieves the pain
- where the patient's current pain is located
- is it a typical VOE
- suggests a different complication,
- advocate for the patient
- assist prescribing clinician to ensure the patient receives adequate analgesia
- work with patients to determine which nonpharmacologic adjunct therapies best relieve the pain.
- educate patient effects of oxygen reduction pain.
- share information on other nonpharmacologic therapies
- include cognitive behavioral therapy, biofeedback, prayer, relaxation techniques, acupuncture, hypnosis, herbal therapies, heat

Treatment

- Evidenced-based Care

Best Practice

- **CLINICAL GUIDELINES APRIL 16, 2020**
- **American Society of Hematology 2020 guidelines for sickle cell disease: prevention, diagnosis, and treatment of cerebrovascular disease in children and adults**

NHLBI Evidenced based Care Guidelines

Hydroxyurea

- March 1998 Hydroxyurea was approved by the U.S. Food and Drug Administration
- patients over 18
- three painful crises in the previous year

Hydroxyurea

The mechanism of hydroxyurea

- **Inhibition of ribonucleotide reductase to inhibit DNA synthesis.**
- **As affect cell devision, cause global myelosuppression including neutropenia, anemia and thrombocytopenia.**

- **Increased HbF production which lead to :**
- Reduce sickling by reduce Hb polymerization.
- RBC lifespan increased
- Improved RBC hydration
- Reduce hemolysis
- Reduce the adhesion of cell to vascular endothelium.

Other mechanism include :

- Increase production of NO by endothelial cell.
- Reduce WBC count and/or neutrophil adhesivity to the vascular endothelium.

Which in general :

- **Reduce frequency/quality of pain**
- **Reduce recurrence of ACS**
- **Reduce hospitalization and need to transfusion**
- **Reduce hemolysis**

Changing world of SCD

Treatments not Universal Cures

- Bone Marrow Transplant
- Gene Therapy
- Stem Cell

New disease-modifying medicines

Endari / L - glutamine

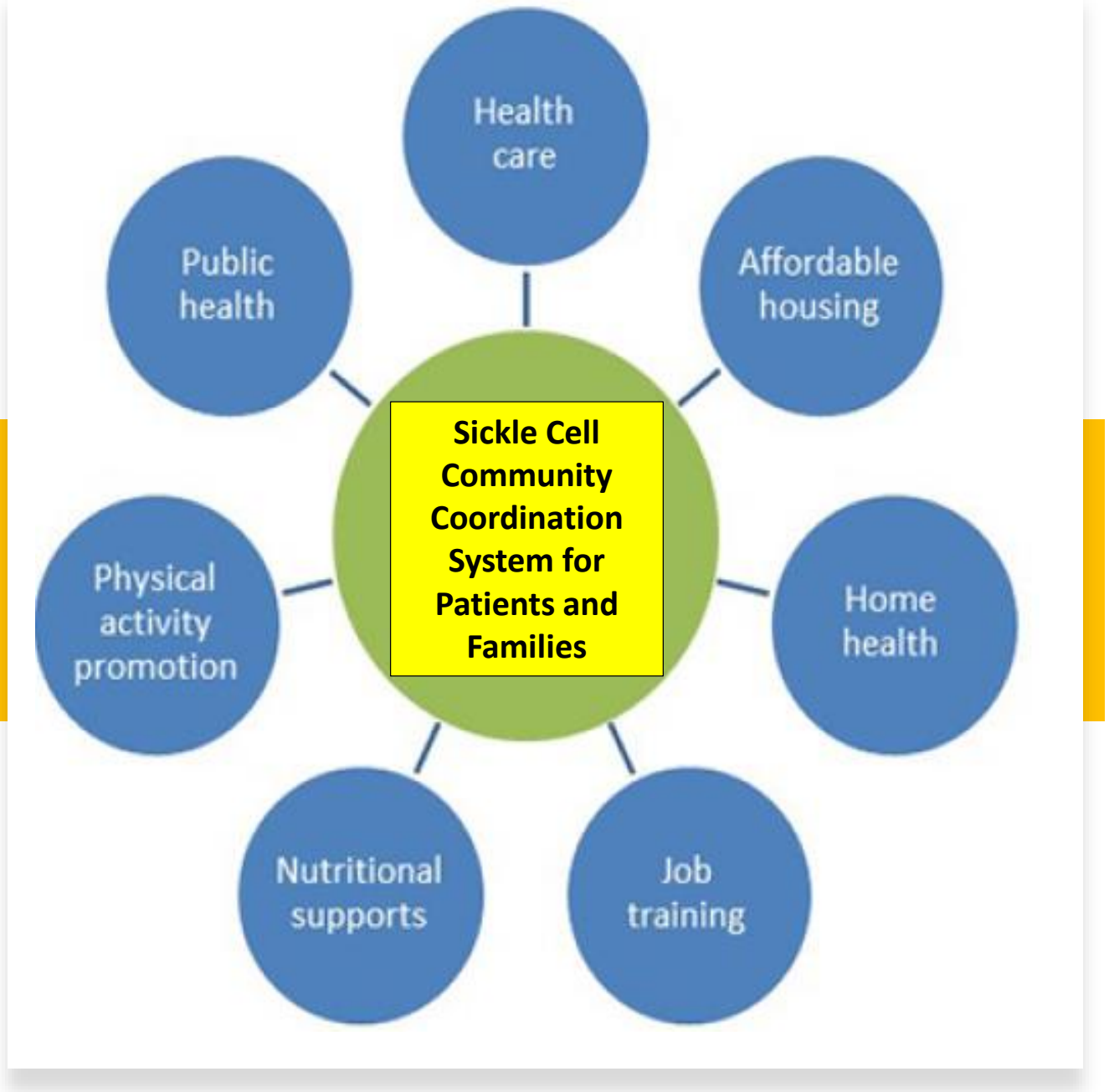
- Pharmaceutical strength
- Action: Prevents oxidative damage to RBC which results in chronic hemolysis and vaso-occlusion (decrease in hospitalizations and time to recovery and better health outcomes

Adakveo (Crizanlizumab)

- infusion
- Action: Reduces frequency of VOC and length of hospitalizations stays
- Children 16 and older and adults

Oxybrita

- Increases hemoglobin affinity – the ability to stick to oxygen
- Blocks polymerization(copying of sickle cells). Potential to modify progression of SCD by lessening damage caused by sickle cells.



Facilitators to care include:

- Knowledgeable consumers and providers
- Awareness and increased use of the NHLBI SCD recommendations
- Appropriate medications for management of symptoms and pain (opioids)
- Patient centered individualized pain protocols
- Use of electronic medical records
- Care-interventions directed at improving provider knowledge and mitigating provider bias

Conclusion

Sickle cell disease (SCD) is a long-term condition that would benefit from a long-term systems of care approach and management.

One that lends with evidence-based best practices, understanding, empathy, and compassion

"Of all forms of discrimination and inequalities, injustice in health is the most shocking and inhuman"

- Martin Luther King. Jr.

For more information contact:
Supporters of Families with Sickle Cell Disease, Inc.
One Organization, Two Locations
Bridging Families One At A Time
918 619 6174
www.sicklecelloklahoma.org

References

- *Mental Health and Learning Needs in Children with Sickle Cell Disease:* Patricia Kavanagh, MD; Assistant Professor of Pediatrics Boston University/Boston Medical Center.

- *Living Well With Sickle Cell In School; An Educational Support and Advocacy Program.* Michael Matthews, Program Director; Children's Sickle Cell Foundation, Inc. Pittsburgh, Pennsylvania , USA

- https://www.nhlbi.nih.gov/

- https://search.usa.gov/search?utf8=%E2%9C%93&affiliate=nhlbi_nhlbi_prod&query=Sickle%20CELL%20&commit=Search

Speaker's Information

Pat L. Corley, RN
Sickle Cell Nurse Educator/Advocate
Cayenne Wellness Center
Axis Advocacy
310.480.9857

Lindsey Corbitt, MA
Community Health Worker
Supporters of Families with Sickle Cell Disease
918. 619. 6174
918. 926.0289

Velvet Brown-Watts, MSW, CM
Executive Director & Parent
Supporters of Families with Sickle Cell Disease, Inc.
918.619.6174
918.926.0100
swithsickelcell@att.net
www.sicklecelloklahoma.org

Shaquanda Lewis,
Patient Advocate
Supporters of Families with Sickle Cell Disease, Inc.
918.619.6174