Transitioning to empowerment

A model for lifespan care of individuals with sickle cell disease

Overview of presentation

- What is meant by the health care transition?
- Models and support of comprehensive care in the US
- An analogy of the current state
- Sickle cell disease: An overview in four slides
- Barriers to a successful transition for individuals with sickle cell disease
- Current models and approaches for facilitation of the transition

What is meant by “health care transition”?

- The process around an individuals transfer of care from a pediatric to an “adult” based system of health care
- Common characteristics of the health care transition
  - Transfer that is driven by chronologic age – 18 to 21 yo
  - A transition process that starts at an earlier chronologic age (12-14 yo)
  - A standardized measurement system
  - Interim incremental outcomes that measure outcomes of the approach
    - Association with some type of curriculum that encompasses aspects of development
    - Often checklists of accomplishment at a given age

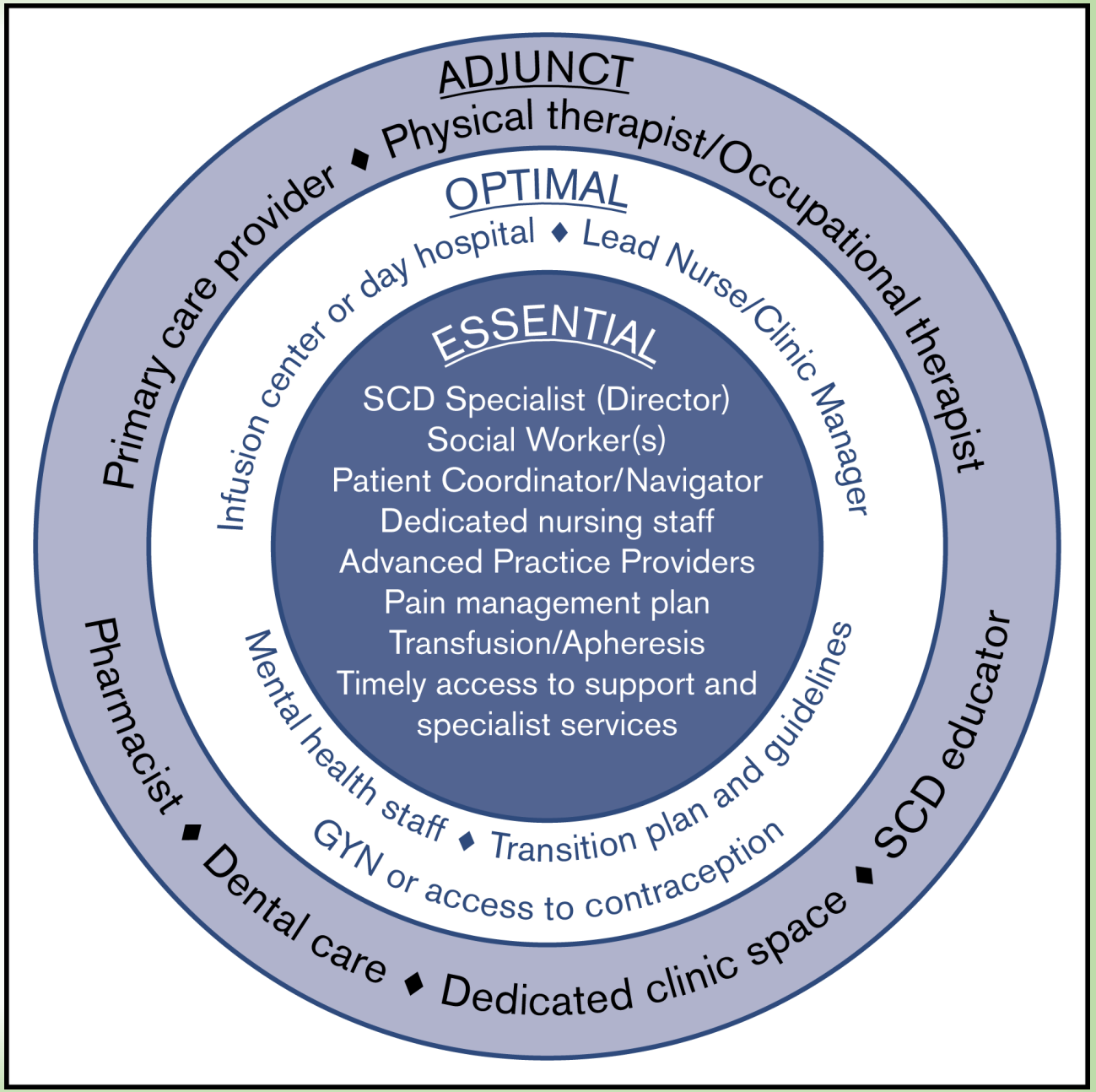
The comprehensive approach to care of inherited disorders: A historical perspective

- Initiated in the 1970s in the US as an approach for longitudinal care of patients with complex, chronic pediatric disorders
- The practice of medicine in the 1970s consisted of :
  - Cures:
    - Sew it up or stabilize it until heals
    - Cut it out (surgical)
    - Kill it
  - Symptom management (bleeding, edema, pain)
  - Limited options for medical management
- Limited options available for multidisciplinary approach at that time

The comprehensive approach to care of inherited disorders: A historical perspective

- Hemophilia and inherited bleeding disorders
  - Hemophilia Treatment Centers funded through Omnibus Reconciliation Act in 1981 through federal grant mechanism within the Maternal and Child Health Bureau
    - Amount of federal funding within the line item has remained largely unchanged since 1980s
  - Expanded to include CDC funding throughout the 1980s (response to HIV)
  - Provides limited funding for support of comprehensive program that included multidisciplinary clinics for the longitudinal care of pediatric and/or adult patients

Components of a SCD comprehensive care clinic



Julie Kanter…Sophie Lanzkron, Building access to care in adult sickle cell disease: defining models of care, essential components, and economic aspects, Blood Adv, 2020.

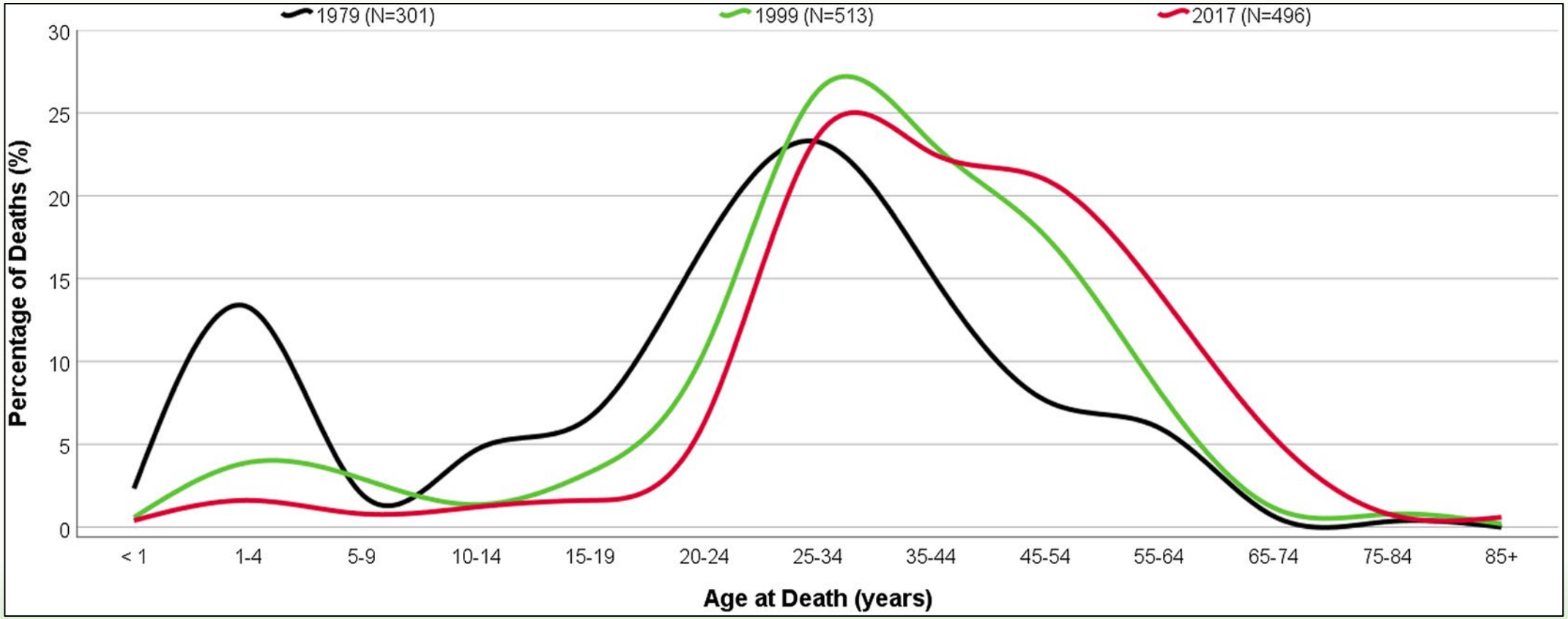
The comprehensive approach to care of inherited disorders: Sustainability

- Financial considerations of a multidisciplinary clinic
- “Without margin {or philanthropic/government financial support}, there is no mission” – source The Web
  - Physical plant – offices, IT, multidisciplinary clinic space
  - Personnel
- Limited options for funding from third party payors – RVU driven
- 340b – Mandatory discounting to selected (by regulation) providers, who then utilize the margin from provider to provide comprehensive services for patients – states typically capture this discount for Medicaid patients
- National Foundation model – Cystic fibrosis
- NIH funding of comprehensive care centers – SCD

Comprehensive care and the transition to adult-based care

- 90-95% of individuals with SCD are alive at the age of 18 y
  - Neonatal screening and identification
  - Early antibiotic prophylaxis and immunization
  - Comprehensive treatment centers

In SCD, a well-managed transition to adult-based care may be a matter of life and death.



Anjelica C…. Hankins, A program of transition to adult care for sickle cell disease, Hematology Am Soc Hematol Educ Program, 2019.

The current model of transition – An analogy

The journey's start

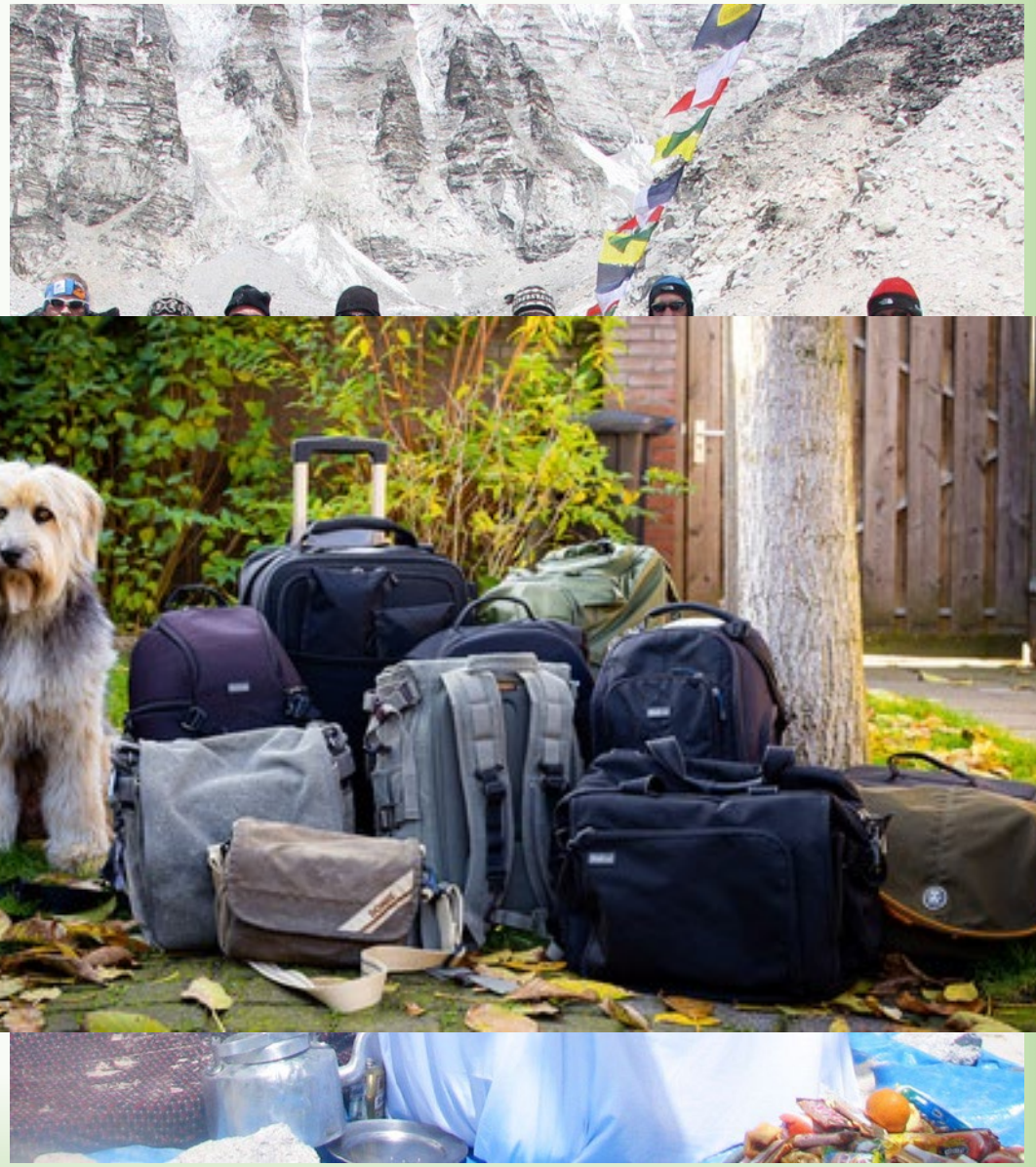
https://www.mhamd.org/what-we-do/outreach-and-education/healthy-new-moms/about-hnm/

https://www.summitclimb.com/page/high-altitude-climbing-sherpas-staff

The transition

The transition

https://www.mhamd.org/what-we-do/outreach-and-education/healthy-new-moms/about-hnm/

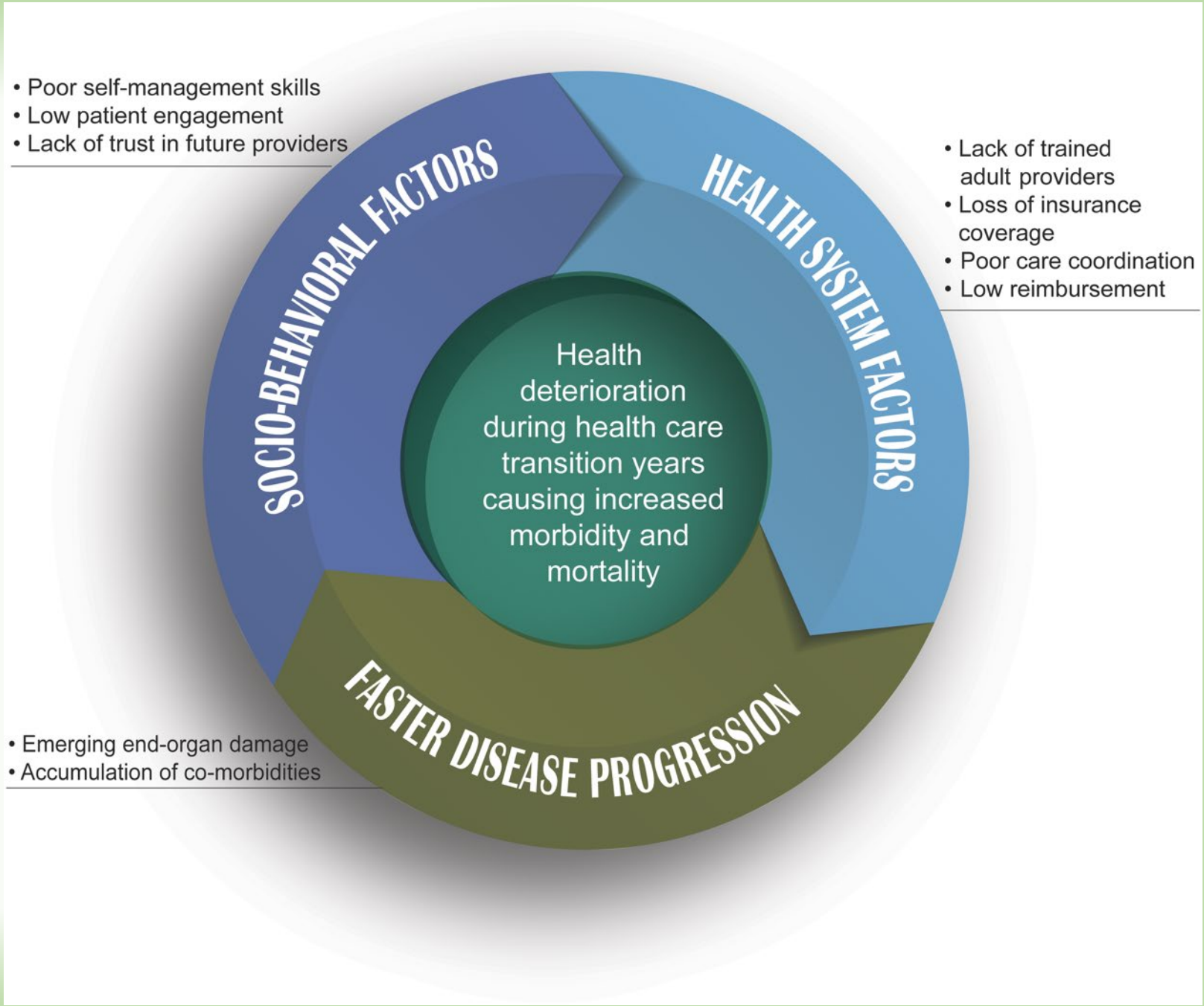


https://www.summitclimb.com/page/high-altitude-climbing-sherpas-staff

The transition

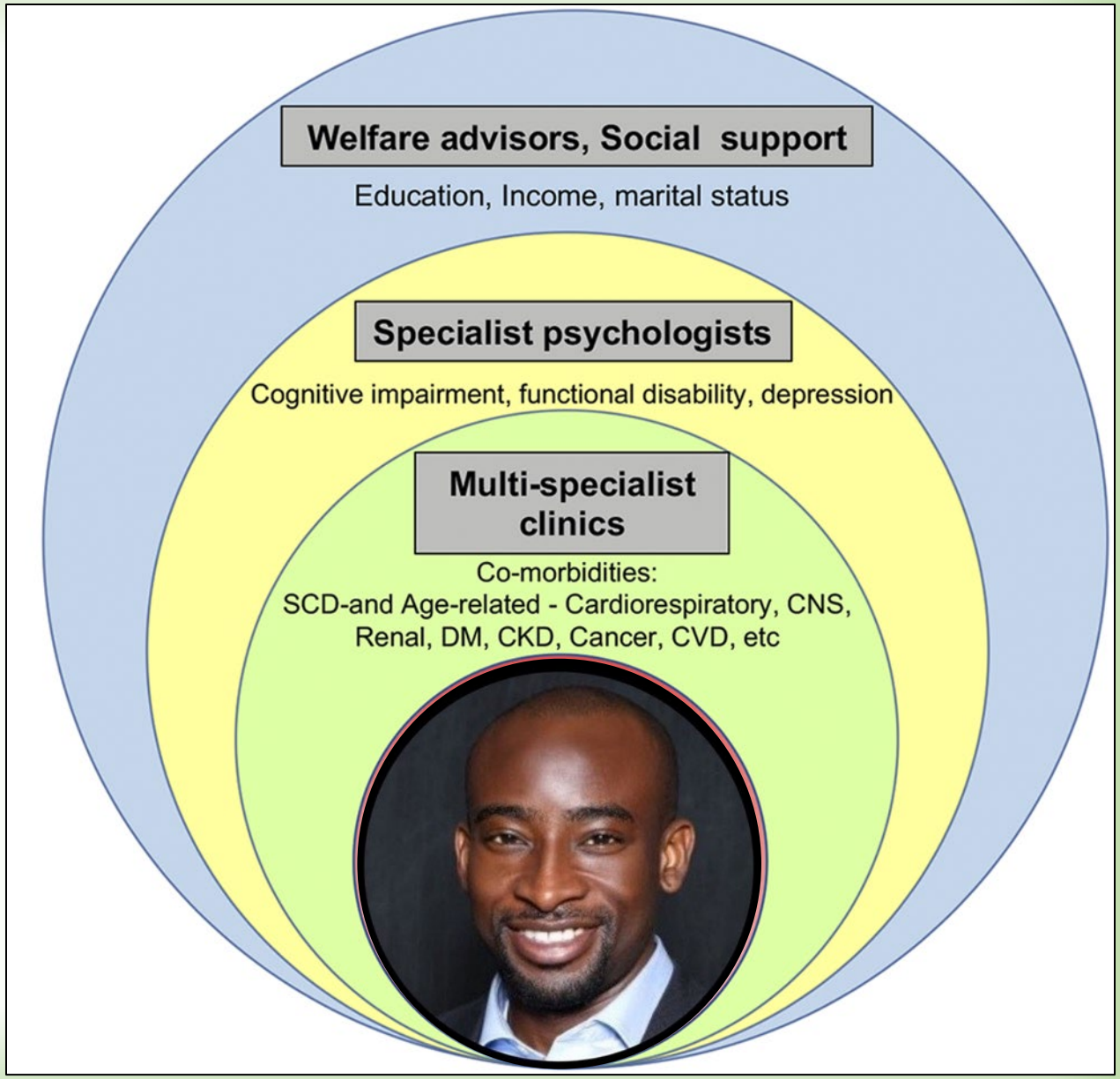
https://external-preview.redd.it/bVjhzbvrw-Z7s09Gu4Oh_4z2EEQRD1j-MYNcJ4bZR3o.jpg?width=640&crop=smart&auto=webp&s=b0edf312222b66a04c5ad634e9a64398c2ac8bc5

Barriers to empowerment



A. Saulsberry...,J. Hankins, A program of transition to adult care for sickle cell disease, Hematology Am Soc Hematol Educ Program, 2019

A patient-centered approach



J.Swee, L.Thein, Optimal disease management and health monitoring in adults with sickle cell disease, Hematology Am Soc Hematol Educ Program, 2019,

https://www.cdc.gov/ncbddd/sicklecell/images/Phillip-Okwo.jpg

How Do We Measure Success?

- High Prescription Volume?
- Low Prescription Volume?
- Number of Admissions?
- Time to Discharge?
- Cost of Care?

These are SYSTEM-Centered Outcomes

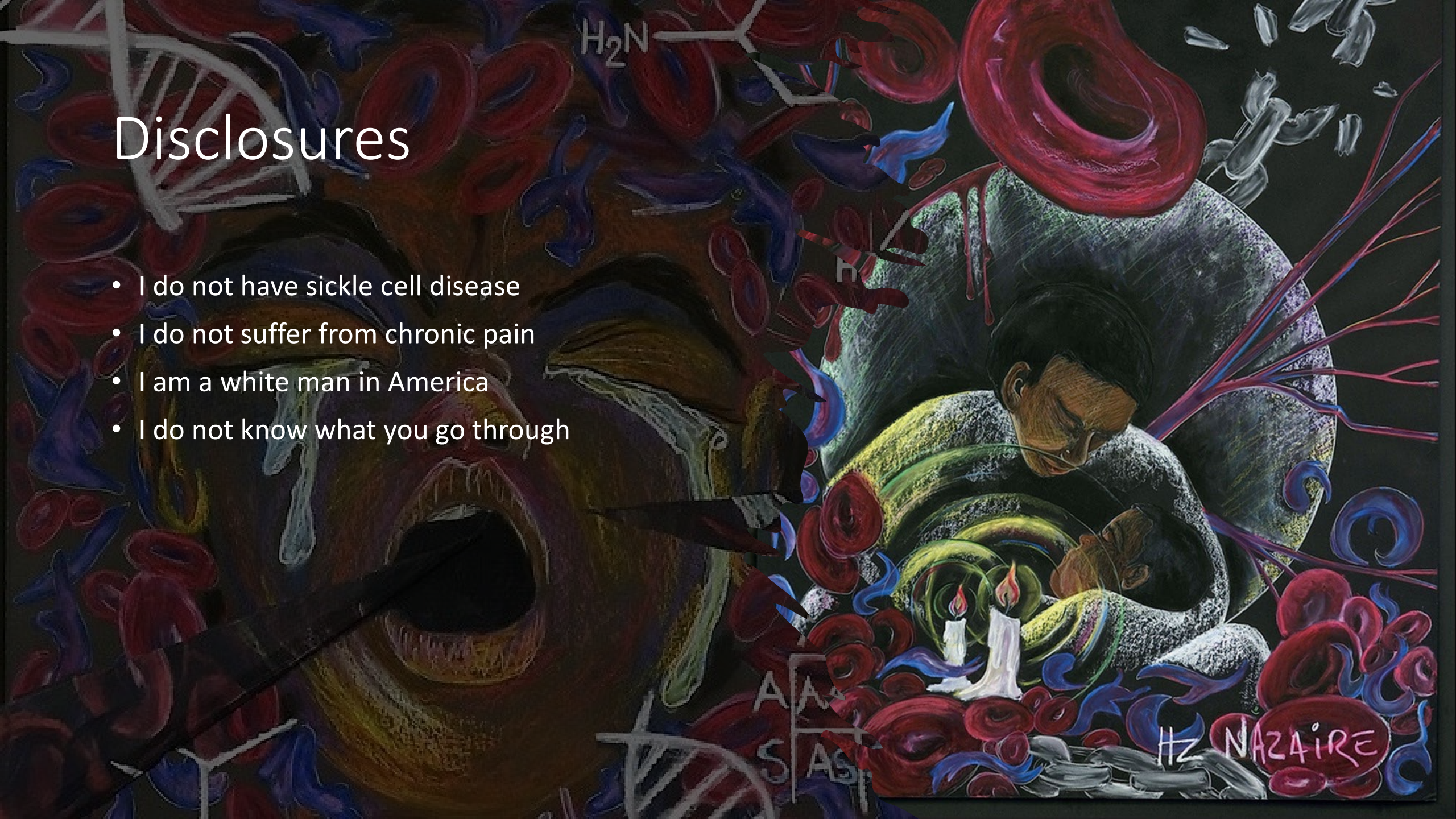
Success MUST be measured by PATIENT-Centered Outcomes

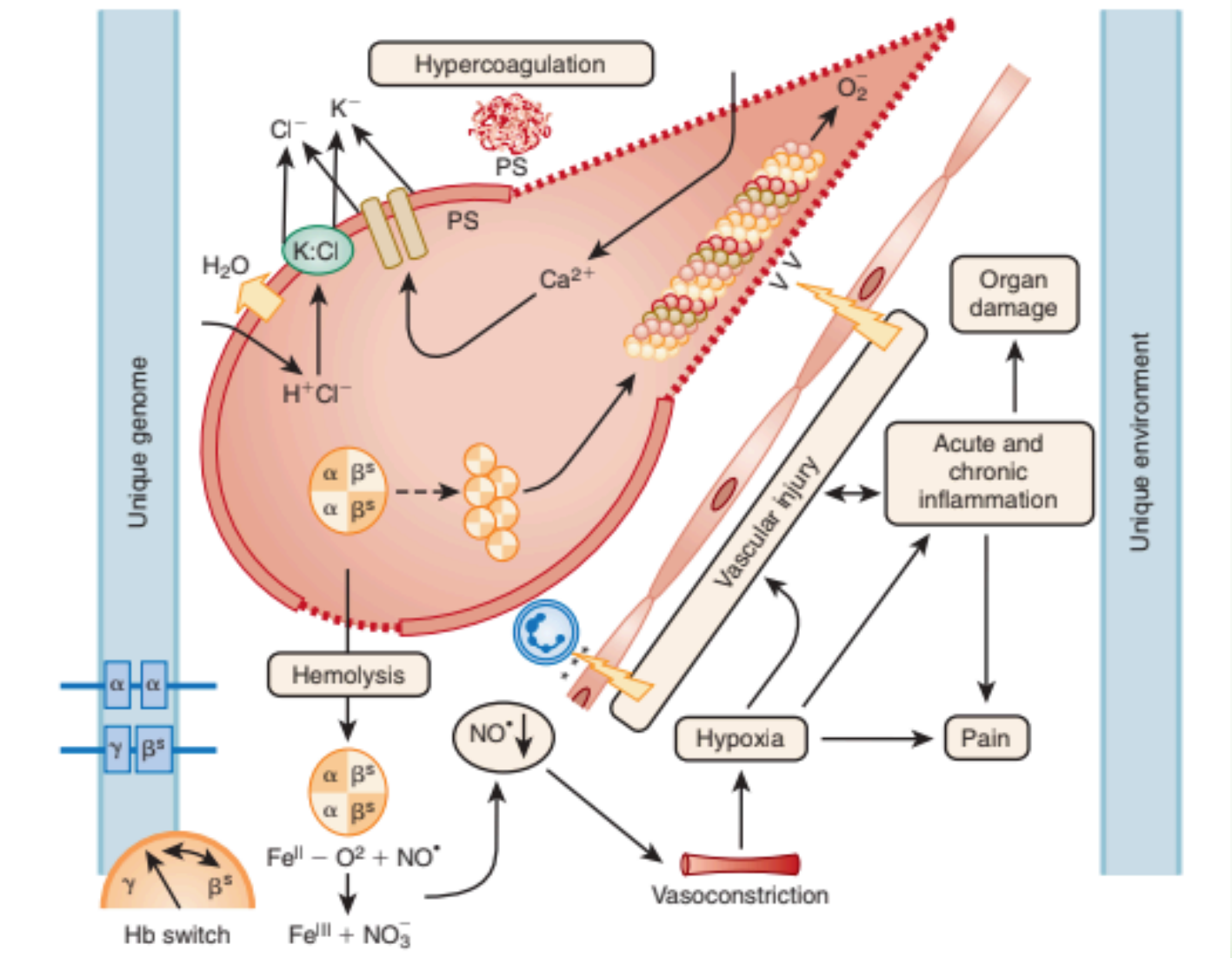
Why Is It Difficult?

- Easier said than done
- We cannot find the solution for better outcomes by looking at the patient
- We find the solution by looking at ourselves

Disclosures

- I do not have sickle cell disease
- I do not suffer from chronic pain
- I am a white man in America
- I do not know what you go through





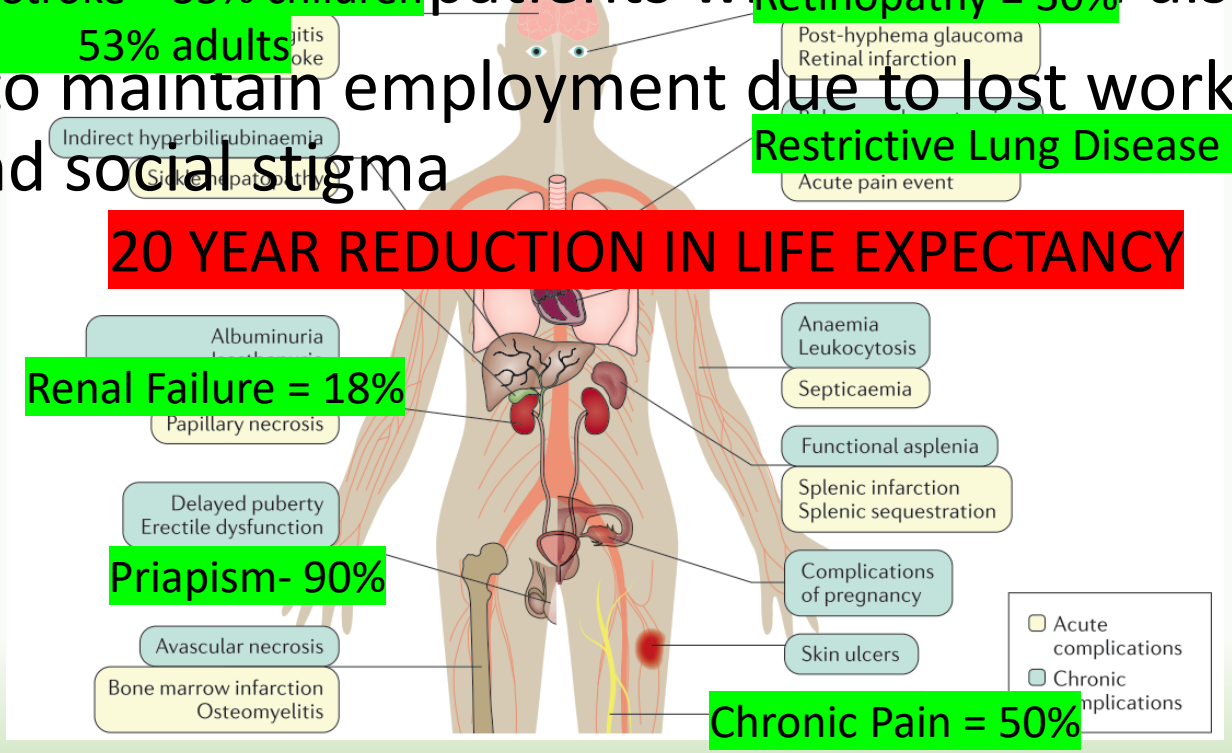
Heeney, Matthew, and Russel Ware. "Sickle Cell Disease." *Nathan and Oski's Hematology of Infancy and Childhood*, by David G. Nathan et al., 8th ed., Elsevier, Saunders, 2015, pp. 676.

Pathophysiology of SCD

- Single mutation in the gene for beta-globin
- Hypoxic or low oxygen conditions lead to changes in RBC and blood vessels
- Hypoxic/Ischemic/reperfusion injury
- Pain
- Silent Damage

A Biopsychosocial Disease

- Multisystemic- Blood is everywhere→Sickling is everywhere
- Depressio[Silent Stroke = 35% children 53% adults] patients wi[Retinopathy = 30%] disease[1]
- Very difficult to maintain employment due to lost workdays, physical limitations, and social stigma



Kato et al. 2017

[1]Adam SS, Flahiff CM, Kamble S, Telen MJ, Reed SD, De Castro LM. Depression, quality of life, and medical resource utilization in sickle cell disease. *Blood Adv*. 2017;1(23):1983-1992. Published 2017 Oct 12. doi:10.1182/bloodadvances.2017006940

Human Disclosures

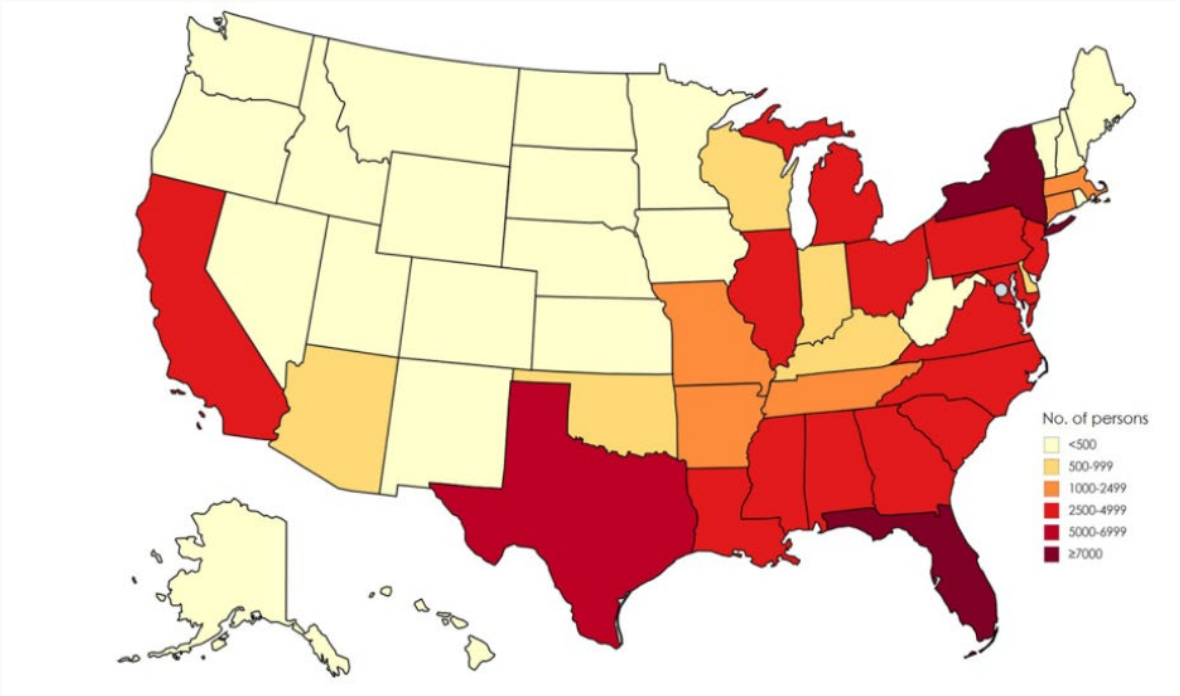
- I do not have sickle cell disease
- I do not suffer from chronic pain
- I am a white man in America
- I do not know what you go through
- I BELIEVE YOU

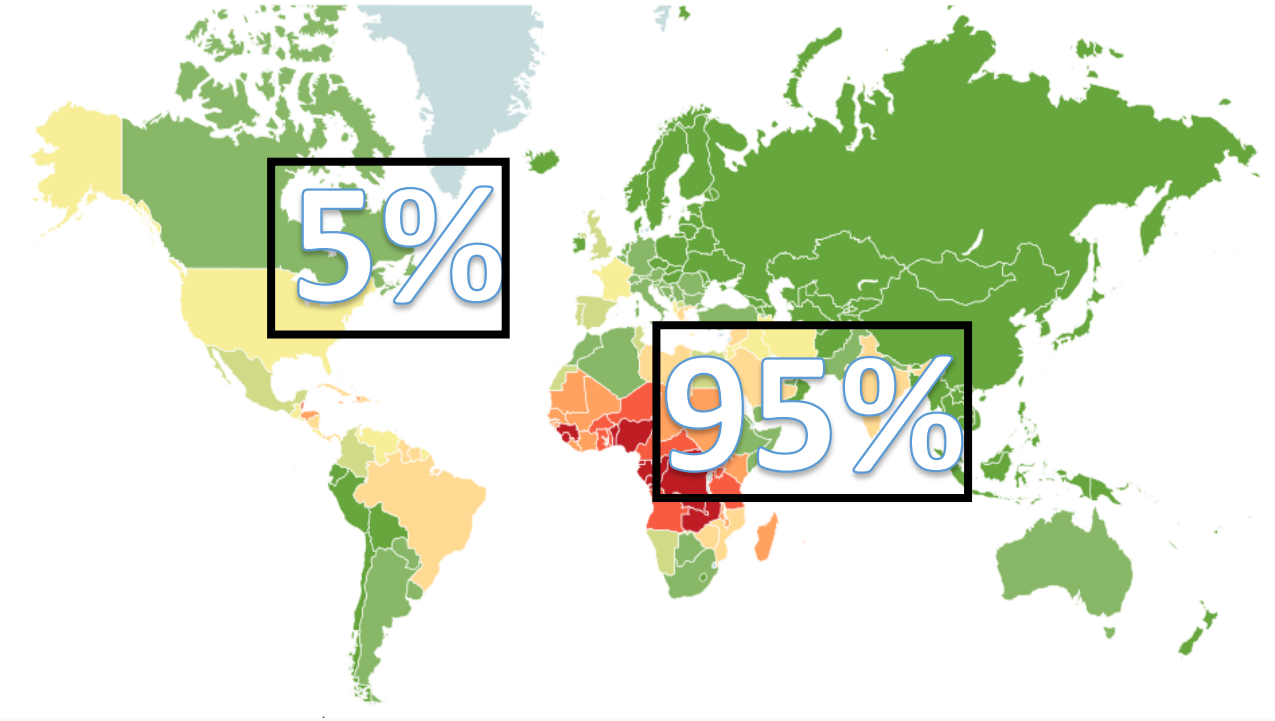
Trust is Earned

- “Respect is an attitude of valuing another person… Trust develops in relationships, in part due to the presence of respect.”

Elander J, Beach MC, Haywood C Jr. Respect, trust, and the management of sickle cell disease pain in hospital: comparative analysis of concern-raising behaviors, preliminary model, and agenda for international collaborative research to inform practice. *Ethn Health*. 2011;16(4-5):405-421. doi:10.1080/13557858.2011.555520

Sickle cell disease: two stories



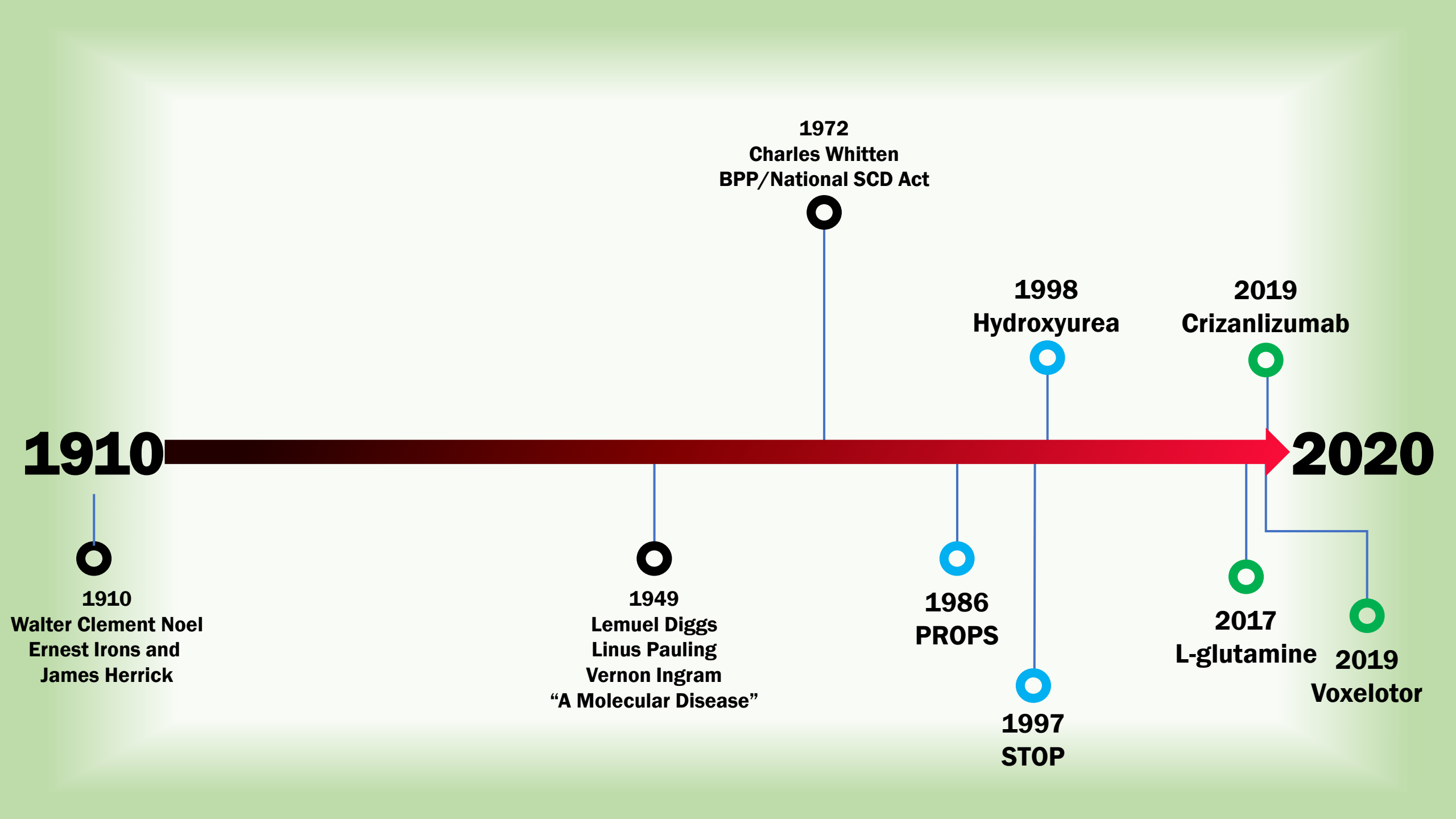


Kato et al 2017
Lee et al 2019

the case of the runaway slave…

“the poor, unfortunate man had leg ulcers and…an absent spleen.”

Lebby, R. Case of Absence of the Spleen.
Southern Journal of Medical Pharmacology (1846)


1910
2020
1910
Walter Clement Noel
Ernest Irons and
James Herrick
1949
Lemuel Diggs
Linus Pauling
Vernon Ingram
“A Molecular Disease”
1972
Charles Whitten
BPP/National SCD Act
1986
PROPS
1997
STOP
1998
Hydroxyurea
2017
L-glutamine
2019
Crizanlizumab
2019
Voxelotor

The Struggle Continues

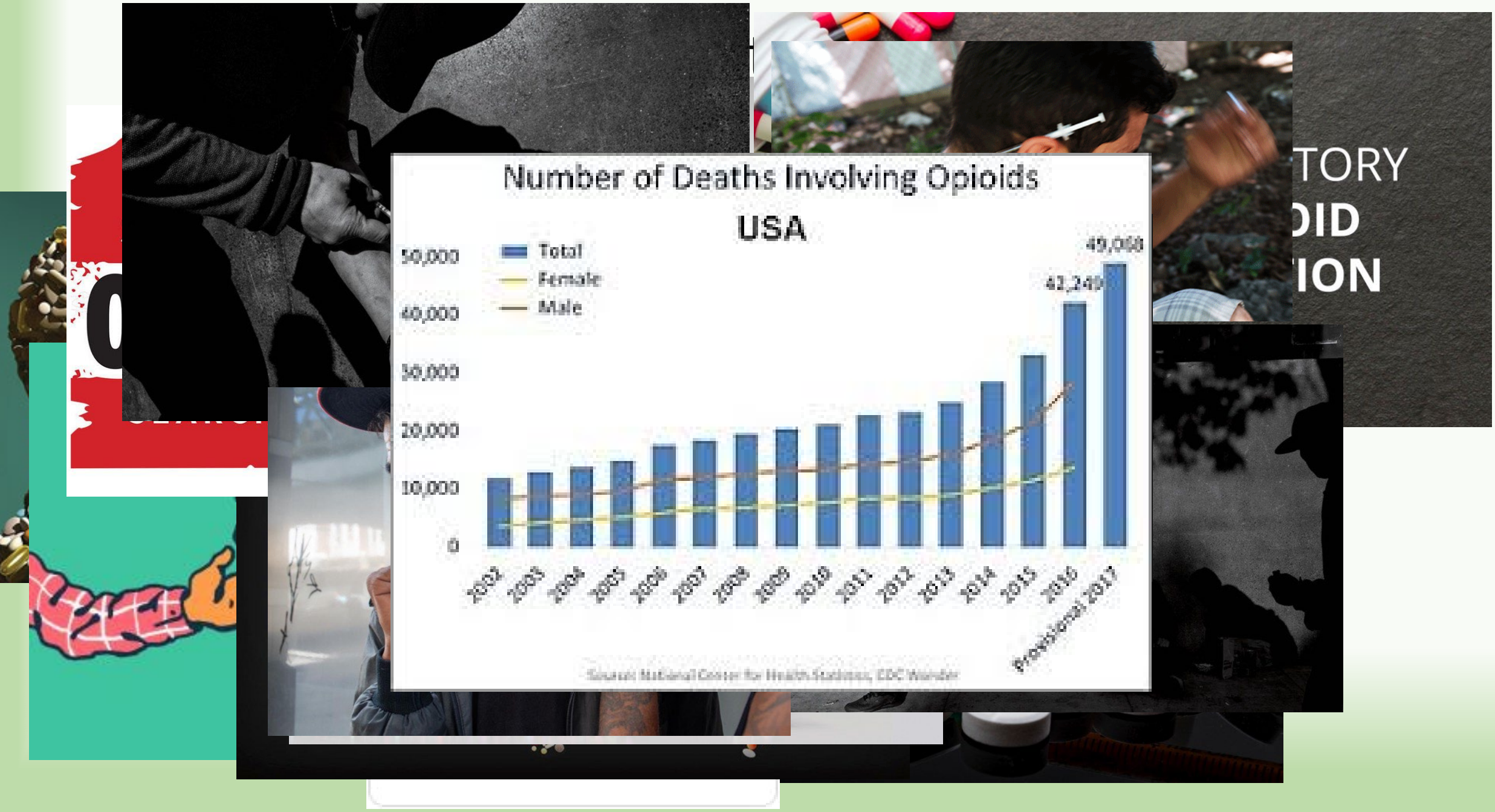
	Cystic Fibrosis	Sickle Cell Disease
U.S. Patient Population	30,000	100,000
NIH Funding per patient	$2807	$812
Foundation Funding	$7690	$102
Registered Clinical Trials	27.3	23.8
Annual Publications	1594	926
Industry Funded Trials	15.6	6.8
Drug Approvals	4	1

Farooq F, Mogayzel PJ, Lanzkron S, Haywood C, Strouse JJ. Comparison of US Federal and Foundation Funding of Research for Sickle Cell Disease and Cystic Fibrosis and Factors Associated With Research Productivity. *JAMA Netw Open.* 2020;3(3):e201737. doi:10.1001/jamanetworkopen.2020.1737

Individual Patient Experiences

- 60% of patients avoid or delay going to ED due to feeling “judged” or ”stigmatized”[1]
- Patients consistently report worse hospital experiences than non-SCD patients[2]

[1]Crego N, Masese R, Bonnabeau E, et al. Patient Perspectives of Sickle Cell Management in the Emergency Department. *Crit Care Nurs Q*. 2021;44(2):160-174. doi:10.1097/CNQ.0000000000000350
[2]Lattimer L, Haywood C Jr, Lanzkron S, Ratanawongsa N, Bediako SM, Beach MC. Problematic hospital experiences among adult patients with sickle cell disease. *J Health Care Poor Underserved*. 2010;21(4):1114-1123. doi:10.1353/hpu.2010.0940


Number of Deaths Involving Opioids
USA
Total
Female
Male
50,000
40,000
30,000
20,000
10,000
0
49,068
42,249
2002
2003
2004
2005
2006
2007
2008
2009
2010
2011
2012
2013
2014
2015
2016
Provisional 2017
TORY
OID
TION

Barriers to Effective Management- Lack of Respect & Trust

- Concern for opioid abuse
- 63% of nurses believed patients with SCD were abusers
  - 30% had concerns about giving SCD patients high doses of opioids
- 53% of ER doctors believed 20% of SCD patients were addicts
- 20% of hematologists believed 20% of SCD patients were addicts

Ruta NS, Ballas SK. The Opioid Drug Epidemic and Sickle Cell Disease: Guilt by Association. Pain Med [Internet]. 2016;17(10):1793–8.

Barriers to Effective Management

- Concern for opioid abuse
- Total 1999-2013:
  - Non-SCD: 174,959
  - SCD: 95

Comparing number of deaths due to opioid pain relievers of non–sickle cell disease patients with the number of deaths due to opioid pain relievers of sickle cell disease patients from 1999 to 2013 in the United States

Year	Non-SCD Patients Who Died Due to OPR	SCD Patients Who Died Due to OPR
1999	4,022	
2000	4,393	
2001	5,521	
2002	7,450	
2003	8,513	
2004	9,856	
2005	10,922	
2006	13,717	
2007	14,401	

Ruta NS, Ballas SK. The Opioid Drug Epidemic and Sickle Cell Disease: Guilt by Association. Pain Med [Internet]. 2016;17(10):1793–8.

Barriers to Effective Management: Lack of System Accountability

The New York Times

TheUpshot

THE NEW HEALTH CARE

A 'Rare Case Where Racial Biases' Protected African-Americans

Fewer opioid prescriptions meant fewer deaths (possibly 14,000), but the episode also reveals how prevalent and harmful stereotypes can be, even if implicit.


By Austin Frakt and Toni Monkovic

Published Nov. 25, 2019 Updated Dec. 2, 2019

Barriers to Effective Management

Among prominent hematology and emergency medicine textbooks “Only 7 texts (37%) note that addiction is infrequent in[the sickle cell population], while 11 (92%) of 12 texts provide such reassurance for cancer-related pain ($P < .005$)

Among 52 ED provider responders (supervising doctors, trainees, nurses:

- 54% felt highly comfortable managing VOC
- Majority not aware of SCD management guidelines existing

Solomon LR. Treatment and prevention of pain due to vaso-occlusive crises in adults with sickle cell disease: An educational void. Blood [Internet]. 2008 Feb 1 [cited 2020 Nov 20];111(3):997–1003.

Martin OY, Thompson SM, Carroll AE, Jacob SA. Emergency Department Provider Survey Regarding Acute Sickle Cell Pain Management. J Pediatr Hematol Oncol [Internet]. 2020 Aug 1 [cited 2020 Nov 10];42(6):375–80.

Not Just Pain

- Pregnancy in women with SCD associated with:
  - IUGR (OR 2.79)
  - Eclampsia (OR 3.02)
  - Perinatal infant mortality (OR 3.76)
  - Peripartum maternal mortality (OR 10.91)
- Similar outcomes in low and high income countries

Boafor TK, Olayemi E, Galadanci N, Hayfron-Benjamin C, Dei-Adomakoh Y, Segbefia C, Kassim AA, Aliyu MH, Galadanci H, Tuuli MG, Rodeghier M, DeBaun MR, Oppong SA. Pregnancy outcomes in women with sickle-cell disease in low and high income countries: a systematic review and meta-analysis. BJOG. 2016 Apr;123(5):691-8. doi: 10.1111/1471-0528.13786. Epub 2015 Dec 15. PMID: 26667608.

Race is Not a Risk Factor

- Multidisciplinary sickle cell obstetric team
- Support and reward system to encourage innovation
- Routine incentive spirometry
- Careful and detailed evaluation/management of all potential VOC and ACS episodes
- Maternal and perinatal mortality reduced by 90%

Oppong SA, Asare EV, Olayemi E, et al. Multidisciplinary care results in similar maternal and perinatal mortality rates for women with and without SCD in a low-resource setting. Am J Hematol. 2019;94:223-230. https://www.ncbi.nlm.nih.gov/pubmed/30456766

Considerations

- Must overcome inertia
  - Patient attitudes
  - Payer assumptions
  - Physician egos

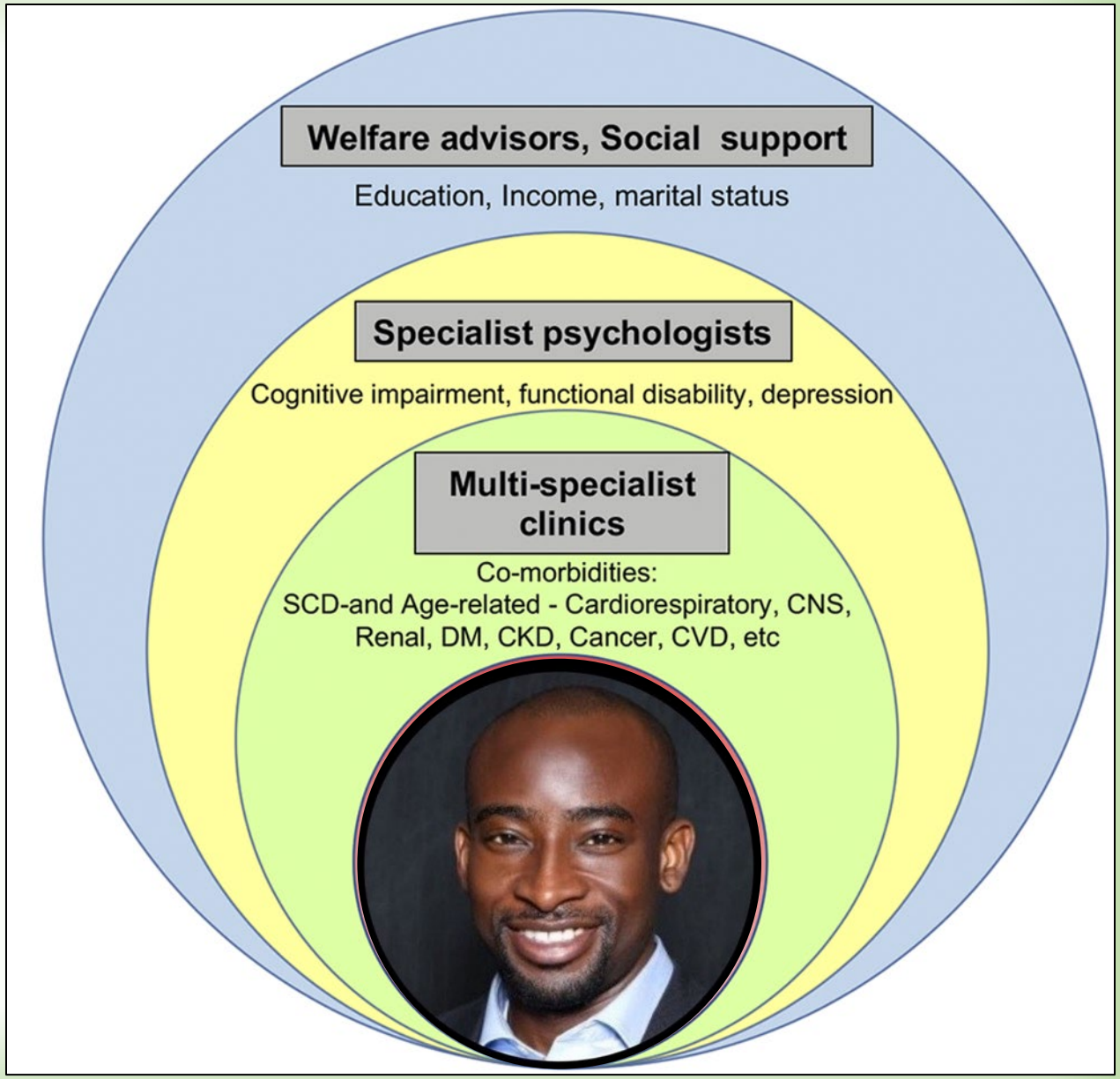
Not Just Pain

- Poor treatment leads to avoidance→Patients lost
- Impossible to navigate systems lead to avoidance→ Patients lost
- Frustrated/angry patients labeled difficult and “fired”→ Patients lost

- System-Centered Solution:
  - Less bounce backs, less opioid prescriptions, fewer time consuming conversations for understaffed hospitals/clinics-
  - Problem Solved
- Patient-Centered Solution
  - Find the Patient. Give them a home.

Lifespan care in Lansing

A community approach to lifespan care



https://www.cdc.gov/ncbddd/sicklecell/images/Phillip-Okwo.jpg