

The Comprehensive Pediatric Sickle Cell Program

World Sickle Cell Day

June 19, 2018



University of
Pittsburgh



Children's | of
Hospital of Pittsburgh | UPMC

Meet the Sickle Cell Team

Providers:

- Margaret Holtz, PA-C:
 - Fall 2014
- Kelsey Platte, PA-C
 - Winter 2016
- Cheryl Hillery, MD
 - Summer 2015
- Amma Owusu-Ansah, MD
 - Summer 2016
- Ram Kalpatthi, MD
 - Spring 2018

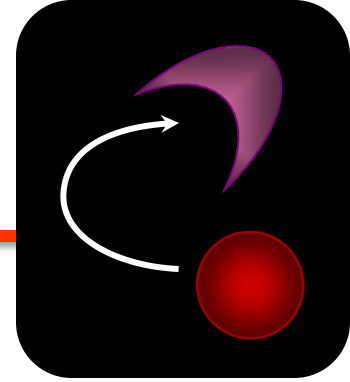
Meet the Sickle Cell Team

- Pamela Mwiinde, MSW
 - 2016: sickle cell social worker
- Janessa Nowlen, PsyD
 - 2017: psychologist/behavioral health
- Suzanne Komaniak, RN, BSN
 - 2017: sickle cell nurse coordinator
- Ashley Kosarich
 - 2015: hematology administrative assistant

Meet the Sickle Cell Team



Our families



~210 children and young adults

- Sickle Cell Disease type:
 - 60% HbSS
 - 30% HbSC
 - 3% HbS β° Thal; 7% HbS β^{+} Thal
 - <1% HbSE; HbS-HPFH
- Age and gender:
 - Age: 0 to 21 yr old
 - 50% male; 50% female

Multidisciplinary care

9C Clinic

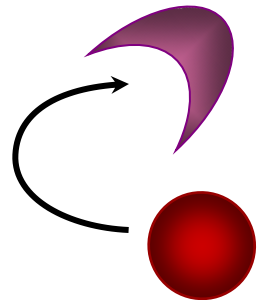
- Clinic and laboratory staff:
 - VS, med check, lab draw, scheduling
- Provider: Maggie, Kelsey, physicians
 - Hx, PE, medical management, patient care plan
- Social worker: Pam Mwiinde
 - Psychosocial assessment, insurance, school
- Nurse coordinator: Suzanne Komaniak
 - Nursing assessment, education, phlebotomy
- Behavioral Health: Dr Janessa Nowlen
 - Sickle cell clinic Tue and Wed
- CSCF and Lend and research people

Pneumococcal infection in sickle cell disease



kayla

- ❖ Newborn screen to identify infants with sickle cell disease at birth
- ❖ Initiate penicillin prophylaxis before the age of 2 to 3 months
- ❖ Treat fever ($T > 101.5^{\circ}\text{F}$) as an emergency
 - Families and medical care providers
- ❖ Pneumococcal immunizations:
 - PSV23 (Pneumovax)
 - PCV7 (Pevnar 7)
 - PCV13 (Pevnar 13)
- ❖ **Stop penicillin when over 6 yo?**



Multidisciplinary care

Stroke prevention

- TCD screening
- Chronic transfusions

School success

- Neurocognitive testing
- Communicate with school : letters, 504 plan, IEP
- Behavioral Health
- School nurse

Multidisciplinary care

Transition

- Work with adult team and CSCF transition program
- Start developing transition skills at a younger age
- 18 yo: new expectations (clinic and hospital)
- Dr Janessa Nowlen and Pam Mwiinde

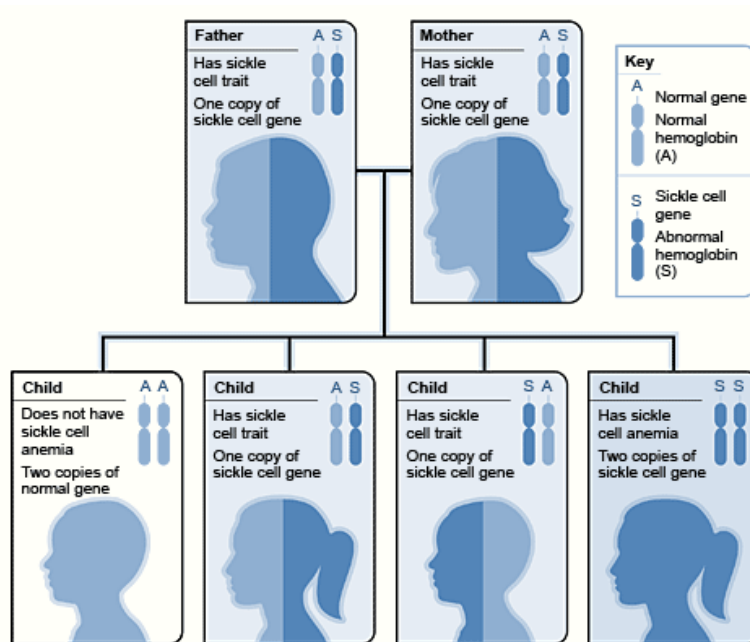
General sickle cell care

- Hydroxyurea
- Sickle cell pain
- Eye (ophthalmology) exams
- Dental exams

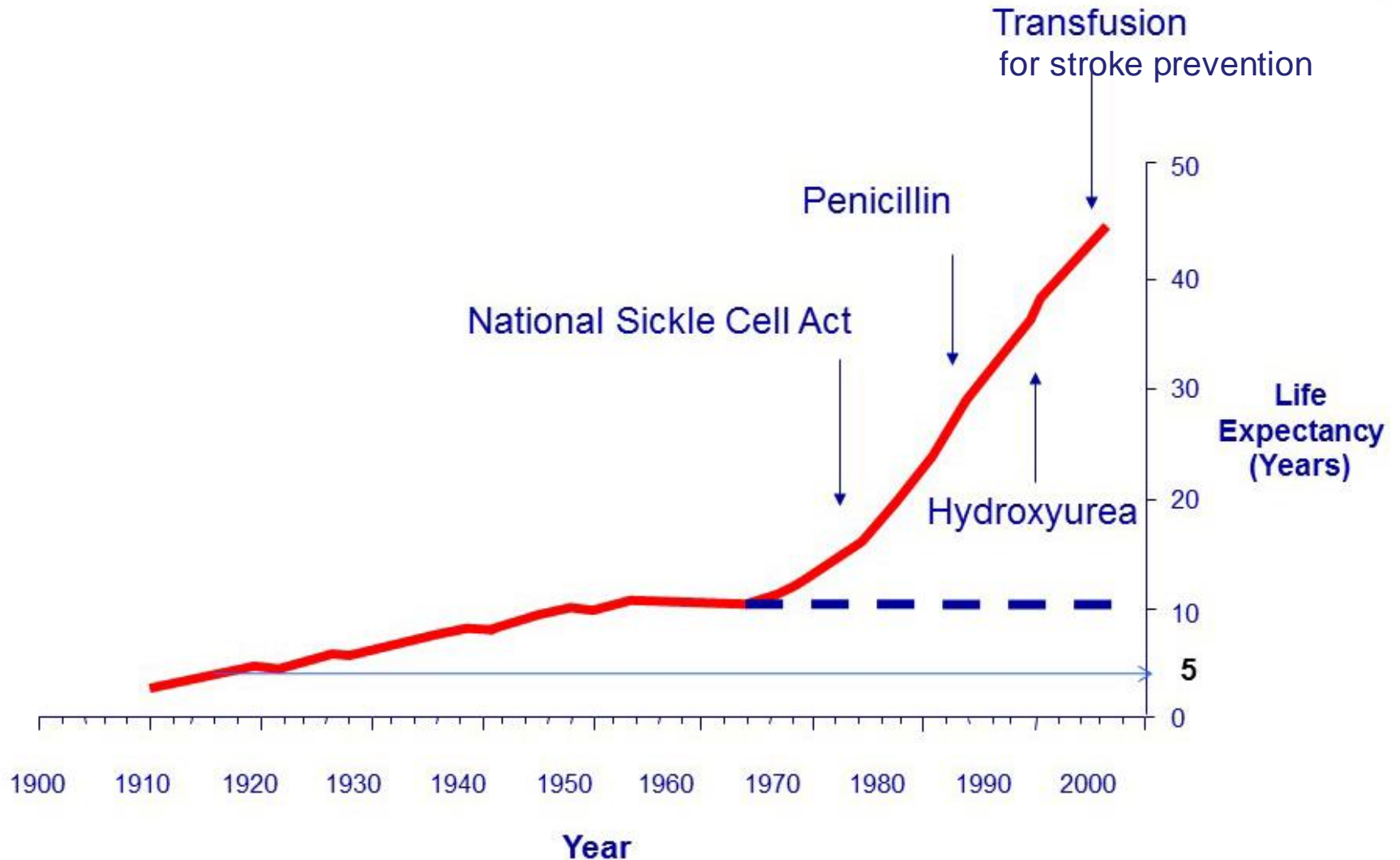
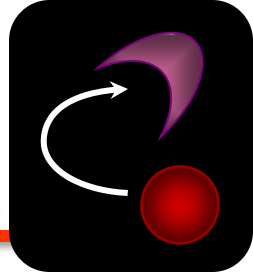
Multidisciplinary care

Genetic counseling:

- Elena Kessler, Sickle cell genetic counselor
- **New this summer:** Tuesday afternoon Clinics
- New families, middle school, teens/transition



Life expectancy in SCA



Education and Outreach



- Kennywood family day/walk
- Summer camp at Flying Horse Farms
- Holiday party
- Social work, genetic and medical trainees in clinic
- LEND program for outreach to families
- Education of school nurses
- Erie Outreach clinic: twice yearly
- Pitt athlete sickle trait education & testing
- CSCF is a major partner!

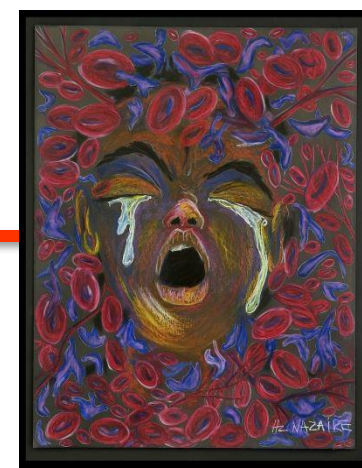
Research initiatives:



Clinical research:

- Sickle cell data registry (CSCF)
- GBT 440 (Global Blood Therapeutics):
 - Anti-sickling agent; outpatient
- RESET/Rivipansel (Pfizer):
 - Anti-adhesion (selectin); inpatient crisis
- *Cure*: bone marrow transplant
 - STRIDE: individuals > 16 yo
 - Alternative donors: cord blood, unrelated, half-match
- OMEG-411 (Sancillo): soon
 - Omega-3 fatty acids; outpatient
- Tissue bank: soon

Research initiatives:



"Ten Redefined" by
Hertz Nazaire

Basic and translational research:

- Novel mechanisms of pain in SCD (Hillery)
- Mechanisms of Platelet activation in SCD (Nolfi)
- Neurocognitive function in SCD (Novelli; Kalpatthi)
- Role of HMGB1 and TLR4 signaling in SCD (Hillery)
- Leukocyte and platelet adhesion in SCD ACS (Sundd)
- Mitochondrial signaling in SCD platelets (Shiva)
- Pulmonary complications of SCD (Gladwin)
- Role of hemin in endothelial dysfunction (Solomon; Kato)

Thanks!



“Hope” by Hertz Nazaire