

Sickle Cell Disease Health Related Quality of Life Documentation in the Electronic Medical Record

L Neumayr, MD, M Treadwell, PhD, P Shah, M Rowland,
K Smith, S Robertson, A Moscicki, MD



100%
kids.



Disclosures

- Grant/Research Support:
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- HRSA Sickle Cell Demonstration Program
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Introduction to Sickle Cell

Normal RBC



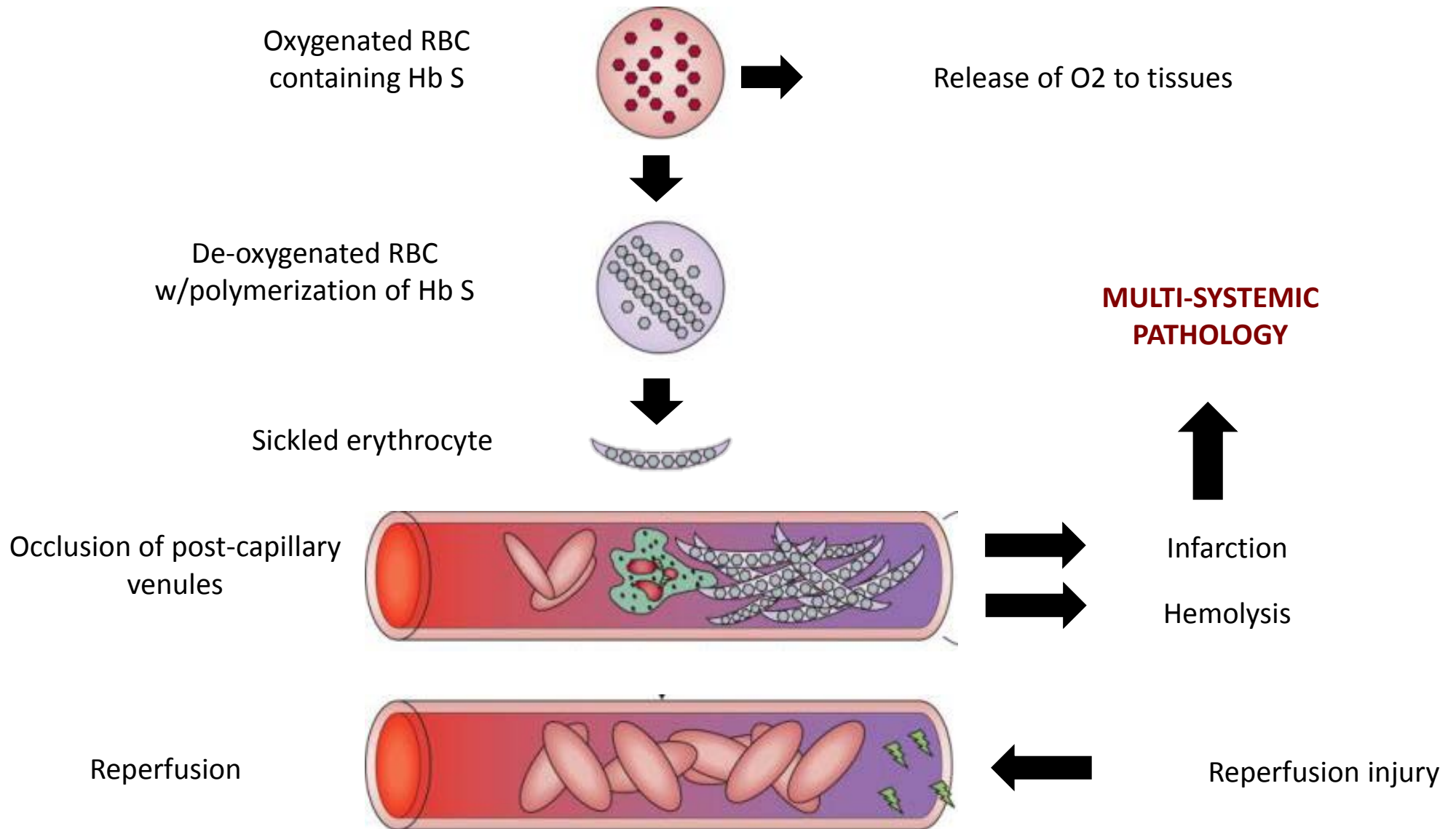
- Round, soft, flexible
- Easily traverse capillary beds
- Lifespan 120 days

Sickle RBC

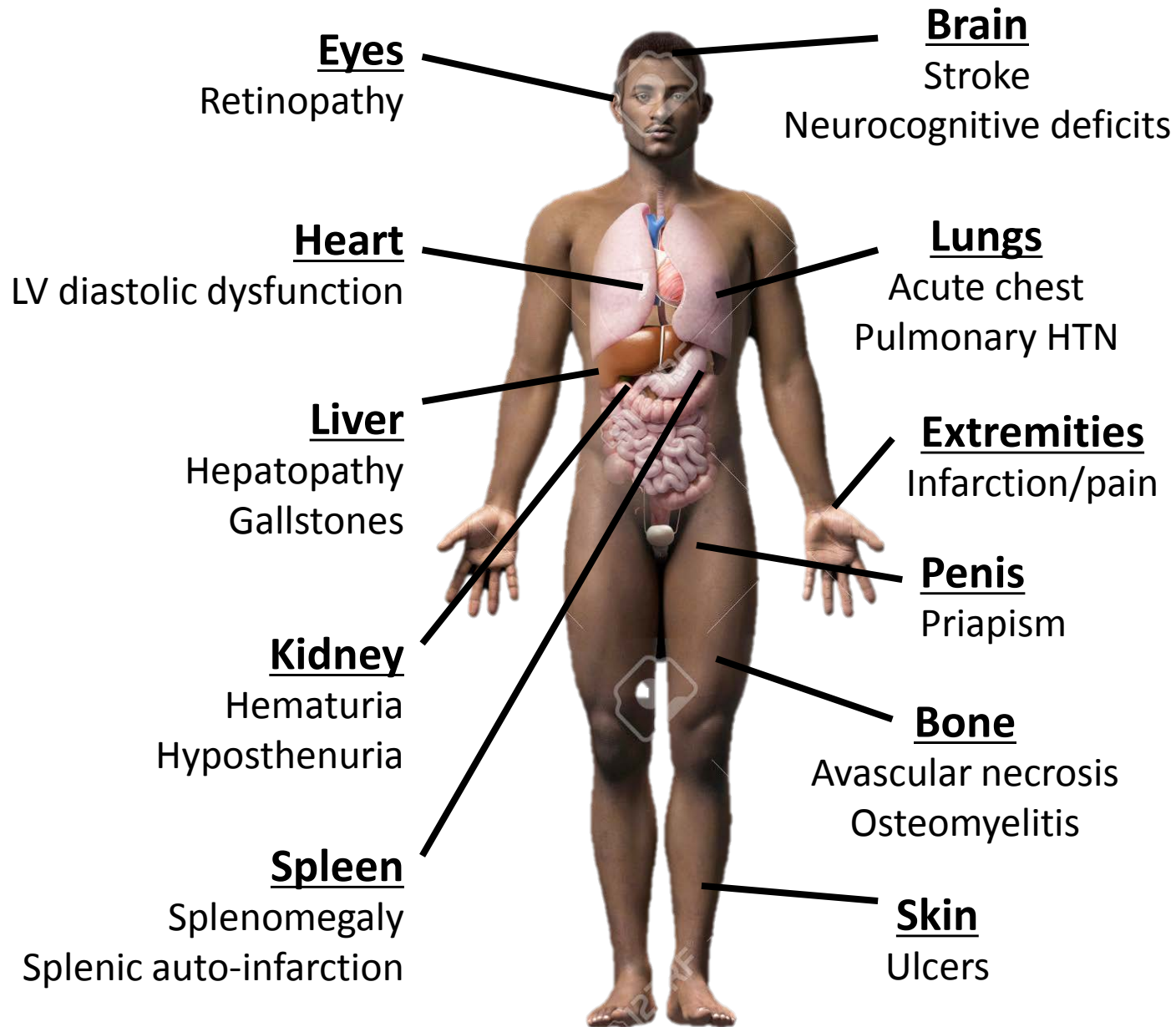


- Rigid, sticky, poorly flexible
- Can clog capillary beds
- Lifespan 7-14 days

Sickle Cell Pathophysiology



Multisystem Pathology in SCD



Current therapy for SCD

- **Hydroxyurea (HU):**
 - Recommended for children with genotypes HbSS or HbSB0
 - ↑ Fetal Hb → ↑ total Hb, ↓ sickling, ↓ pain crises
 - ↓ white cell count, ↓ platelet count → ↓ vaso-occlusion
 - Nitric oxide donor
- Prophylactic penicillin in pediatrics
- Transfusion
- Pain management
- Transplantation, gene therapy

HRQOL in SCD

- HRQOL inventories are common outcome measures in research in sickle cell
- Evaluating patient centered outcomes, specifically HRQOL, in relation to therapeutic interventions in the clinical setting is critical
- Implementation and utilization of HRQOL measures within the electronic medical record (EMR) are challenging

Objectives and Methods

- The PROMIS Patient Global Health (PGH-7) was incorporated into the Epic EMR at UCSF Benioff Children's Hospitals
- The PGH -7 composite score (0-35) measures general, physical, mental and social health
 - PGH-7 is brief, internally consistent, reliable, correlates with other PROMIS measures and PedsQL (Forrest et. al. 2014, 2015)
- Written consent was obtained from patients age 8+ and their families and a paper version of PGH-7 was administered
- Scores were entered into an Epic EMR flow-sheet by a physician after the clinic visit

Screen Shot of PHG-7

Epic Patient Lookup Patient Station In Basket My Reports Remind Me ED Referral OR Schedule OR Reports Appts Beacon Admin Personalize Help Guides Open Case

Male, y.o., Patient Aliases (AKAs): None Race: Primary Ins.: CCS-CA Allergies: No Known Allergies PCP: Marsh, Anne Pref Language: English MyChart: No proxy exists My Sticky Note: Patient Messages: None Difficult Airway: None Last Ht: 131.5 cm Last Wt: 36 kg Lst BSA: 1.13 m2 MARSH, A Unit: PEDSC

CSN: Advance Directive Filed?: None Outside Info: **New Out 1...**

Special Assessments

HARMONY QOL **PROMIS PGH-7+2**

HARMONY QOL
New Reading

No data found.

PROMIS PGH-7+2 - XUC PROMIS PGH-7+2
Time taken: 20 / 2017

Responsible Create Note

PROMIS Pediatric Global Health-7+2 © 2008-2012 PROMIS Health Organization and PROMIS Cooperative Group

Please respond to each item by marking one box per row.

In general,

Would you say your health is:	Excellent	Very good	Good	Fair	Poor
Would you say your quality of life is:	Excellent	Very good	Good	Fair	Poor
How would you rate your physical health?	Excellent	Very good	Good	Fair	Poor
How would you rate your mental health, including your mood and your ability to think?	Excellent	Very good	Good	Fair	Poor
How often do you feel really sad?	Never	Rarely	Sometimes	Often	Always
How often do you have fun with your friends?	Never	Rarely	Sometimes	Often	Always
How often do your parents listen to your ideas?	Never	Rarely	Sometimes	Often	Always

PGH-7+2. In the past seven days,

I got tired easily.	Never	Rarely	Sometimes	Often	Always
I had trouble sleeping	Never	Rarely	Sometimes	Often	Always

Results

- 66 patients with HbSS or HbSBeta0, 33 with other genotypes
- Avg age: 13.5 ± 4.1, 50 females, 49 males
- Genotype, gender, annual family income categories were not predictors of PGH-7 scores in SCD
- 61% HbSS/HSB0 prescribed HU did not have higher PGH-7 scores

	SCD (n=99)	Controls (n=3,635)* (25% with chronic disease)	P value
Total PGH-7 Score	26.3 ± 4.7	28.7 ± 4.7	< .001
PGH-7 Mental Health Item	4.0 ± 1.2	4.2 ± 1.0	ns
PGH-7 Physical Health Item	3.5 ± 1.1	4.2 ± 0.9	< .001

* Forrest et al, Qual of Life Res 2014

Conclusions

- We demonstrated the feasibility of building the PGH-7 questionnaire into an Epic EMR
- A flow-sheet format can lead to tracking of scores over time and reports
- Patients and families were interested in HRQOL
- This preliminary data show slightly lower scores in pediatric patients with SCD but did not identify predictors of PGH-7

Limitations

- PGH-7 not validated in SCD
- Small, convenience sample size
- No longitudinal data collected yet
- Comparison group retrospective; much larger cohort

Future Directions

- Study approved by IRB for oral consent
- Real-time entry of PGH-7 data during clinic visits
- Longitudinal PGH-7 data collection
- Build Epic report for summary of longitudinal HRQOL and clinical data → improved tracking of impact of HU dose, compliance and hematologic parameters in comparison to PGH-7 scores
- PGH-7 data collection for SCD patients on chronic transfusion
- Patient/family access to PGH-7 questionnaire via MyChart

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- Epic Builders at UCSF Benioff Children's Hospitals