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

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- Site PI, Pfizer Pharmaceutical Co.
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
Oxygenated RBC containing Hb S

De-oxygenated RBC w/polymerization of Hb S

Sickled erythrocyte

Release of O2 to tissues

Occlusion of post-capillary venules

MULTI-SYSTEMIC PATHOLOGY

Infarction

Hemolysis

Reperfusion

Reperfusion injury
Multisystem Pathology in SCD

**Eyes**
- Retinopathy

**Brain**
- Stroke
- Neurocognitive deficits

**Heart**
- LV diastolic dysfunction

**Lungs**
- Acute chest
- Pulmonary HTN

**Liver**
- Hepatopathy
- Gallstones

**Kidney**
- Hematuria
- Hyposthenuria

**Spleen**
- Splenomegaly
- Splenic auto-infarction

**Penis**
- Priapism

**Bone**
- Avascular necrosis
- Osteomyelitis

**Skin**
- Ulcers

**Extremities**
- Infarction/pain

**Multisystem Pathology in SCD**

- Brain
  - Stroke
  - Neurocognitive deficits

- Lungs
  - Acute chest
  - Pulmonary HTN

- Liver
  - Hepatopathy
  - Gallstones

- Kidney
  - Hematuria
  - Hyposthenuria

- Spleen
  - Splenomegaly
  - Splenic auto-infarction

- Penis
  - Priapism

- Bone
  - Avascular necrosis
  - Osteomyelitis

- Skin
  - Ulcers
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

**Special Assessments**

**HARMONY QOL**

- **PROMIS PGH-7+2**
- **New Reading**

No data found.

**PROMIS PGH-7+2 - XUC PROMIS PGH-7+2**

- **Time taken:** 20 s 1/22/2017

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

**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

<table>
<thead>
<tr>
<th></th>
<th>SCD (n=99)</th>
<th>Controls (n=3,635)* (25% with chronic disease)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total PGH-7 Score</td>
<td>26.3 ± 4.7</td>
<td>28.7 ± 4.7</td>
<td>&lt; .001</td>
</tr>
<tr>
<td>PGH-7 Mental Health Item</td>
<td>4.0 ± 1.2</td>
<td>4.2 ± 1.0</td>
<td>ns</td>
</tr>
<tr>
<td>PGH-7 Physical Health Item</td>
<td>3.5 ± 1.1</td>
<td>4.2 ± 0.9</td>
<td>&lt; .001</td>
</tr>
</tbody>
</table>

* 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
Acknowledgements

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- Epic Builders at UCSF Benioff Children’s Hospitals