



## The 65th ASH Annual Meeting Abstracts

## POSTER ABSTRACTS

## 904. OUTCOMES RESEARCH-NON-MALIGNANT CONDITIONS

**Illness Perception and Stigma Among Children and Adolescents with Sickle Cell Disease**Savannah Winkler, BSc<sup>1</sup>, Kathryn King, BSc<sup>1</sup>, Robert I. Liem, MD<sup>2</sup>, Jane Holl, MD<sup>3</sup>, Sherif M. Badawy, MD MBA, MS<sup>1,2</sup><sup>1</sup>Division of Hematology, Oncology, Neuro-Oncology and Stem Cell Transplant, Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, IL<sup>2</sup>Department of Pediatrics, Northwestern University Feinberg School of Medicine, Chicago, IL<sup>3</sup>Department of Neurology and Center for Healthcare Delivery Science and Innovation, University of Chicago, Chicago, IL**Background:**

Stigma and illness perceptions surrounding adolescents and young adults (AYA) with sickle cell disease (SCD) are not well-studied. The psycho-social and emotional factors may have implications on overall patient-reported outcomes, including health-related quality of life (HRQOL) and medication adherence behavior.

**Aims:**

- (1) Evaluate the impact of patients' perceptions of SCD and their medications on HRQOL and adherence
- (2) Examine the relationship of illness perception and stigma among AYA with SCD

**Methods:**

In this longitudinal, single-institution study, 68 participants were enrolled from outpatient hematology clinic at Lurie Children's Hospital of Chicago. Patients were approached if they were between 10 to 25 years old, English-speaking, and taking hydroxyurea, voxelotor and/or crizanlizumab for SCD treatment. Participants completed the Brief-Illness Perception Questionnaire (B-IPQ), Patient Reported Outcomes Measurement Information System (PROMIS) HRQOL Domains, Measure of Sickle Cell Stigma Questionnaire (MoSCS), Visual Analogue Scale (VAS) and Adherence to Refills and Medications Scale 7 (ARMS-7) every 3-4 months. Patient statistics were evaluated utilizing Wilcoxon (Mann-Whitney) rank sum tests, Spearman rho correlations ( $r_s$ ), and chi-square analysis. The data presented in this abstract represent baseline analysis for all enrolled, active study participants.

**Results:**

Sixty-eight participants completed study assessments (90% Black; 53% Female) with a median age of 14 years old (IQR 11-17) and 84% HbSS, homozygous hemoglobin S. In Table 1, patients with less perceived personal control of SCD and more emotional response to SCD reported worse social isolation ( $r_s = -0.32$ ,  $p = 0.01$ ;  $r_s = 0.31$ ,  $p < 0.01$ ), mobility ( $r_s = 0.34$ ,  $p < 0.01$ ;  $r_s = -0.27$ ,  $p = 0.03$ ), anxiety ( $r_s = -0.29$ ,  $p = 0.02$ ;  $r_s = 0.38$ ,  $p < 0.01$ ), depression ( $r_s = -0.29$ ,  $p = 0.02$ ;  $r_s = 0.42$ ,  $p < 0.001$ ), and fatigue ( $r_s = -0.41$ ,  $p < 0.001$ ;  $r_s = 0.3$ ,  $p < 0.01$ ), respectively. Further, more perceived personal control was associated with less physical stress ( $r_s = -0.49$ ,  $p < 0.001$ ), pain ( $r_s = -0.26$ ,  $p = 0.03$ ), fatigue ( $r_s = -0.41$ ,  $p < 0.001$ ), and greater mobility ( $r_s = 0.34$ ,  $p < 0.01$ ). Participants with more negative perceptions of SCD consequences and concerns reported more physical stress ( $r_s = 0.29$ ,  $p = 0.02$ ;  $r_s = 0.27$ ,  $p = 0.03$ ), depression ( $r_s = 0.32$ ,  $p < 0.01$ ;  $r_s = 0.40$ ,  $p = 0.001$ ), and fatigue ( $r_s = 0.33$ ,  $p = 0.01$ ;  $r_s = 0.37$ ,  $p < 0.01$ ) (Table 1). Additionally, the B-IPQ correlating with PROMIS baseline data indicated patients who had greater understanding of their SCD reported better self-efficacy in both managing their medication and symptoms ( $r_s = 0.29$ ,  $p = 0.01$ ;  $r_s = 0.29$ ,  $p = 0.02$ ). In Table 2, patients with higher self-reported medication adherence using VAS, had better treatment control (5 vs 4,  $p < 0.001$ ) and less perceived SCD-related symptoms (2 vs 3, 0.03). Utilizing ARMS-7, patients with higher self-reported medication adherence perceived less SCD-related consequences (2 vs 3,  $p < 0.01$ ), symptoms (5 vs 4,  $p = 0.01$ ), displayed better treatment control (10 vs 8,  $p = 0.03$ ), but greater emotional response (1 vs 3,  $p = 0.04$ ) (Table 2). Most patients (82%) endorsed  $\geq 1$  statement related to stigma. Patients frequently indicated that they are very careful who they tell they have SCD (64%), telling someone they have SCD is risky (38%), no one knows that they have SCD (52%), and they worry about people discriminating against them because they have SCD (31%). Patients with greater levels of stigma reported worse social isolation ( $r_s = 0.38$ ,  $p < 0.01$ ) and self-efficacy managing symptoms ( $r_s = -0.30$ ,  $p = 0.01$ ).

**Summary/Conclusion:**

The multifaceted relationship between medication adherence, illness perception and stigma related to HRQOL in adolescents and young adults relies upon patients' perceived control over their illness. Future evaluation of influential factors involving patients' perceived control are needed to improve SCD-clinical outcomes.

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**Table 1: Patients' perceptions of SCD and medication in relation to their HRQOL using PROMIS®<sup>1,2,3</sup>**

B-IPQ Domains							
HRQOL Domains	Consequences	Personal Control	Treatment Control	Identity	Concerns	Understanding	Emotional Response
Physical Domains, <i>r<sub>s</sub></i> ( <i>P</i> value)							
Physical Stress	0.29 (0.02)	-0.49 ( <b>&lt;0.001</b> )	-0.09 (0.49)	0.25 (0.04)	0.27 (0.03)	-0.09 (0.43)	0.23 (0.06)
Fatigue	0.33 (0.01)	-0.41 ( <b>&lt;0.001</b> )	0.07 (0.58)	0.31 (0.01)	0.37 ( <b>&lt;0.01</b> )	-0.06 (0.61)	0.33 ( <b>&lt;0.01</b> )
Mobility	-0.18 (0.13)	0.34 ( <b>&lt;0.01</b> )	0.15 (0.23)	-0.14 (0.27)	-0.17 (0.16)	0.10 (0.39)	-0.27 (0.03)
Pain	0.21 (0.09)	-0.26 (0.03)	-0.11 (0.38)	0.27 (0.03)	0.04 (0.74)	-0.09 (0.49)	0.14 (0.24)
Psychological Domains, <i>r<sub>s</sub></i> ( <i>P</i> value)							
Psychological Stress	0.16 (0.19)	-0.23 (0.07)	-0.002 (0.99)	0.12 (0.31)	0.35 ( <b>&lt;0.01</b> )	-0.09 (0.47)	0.50 ( <b>&lt;0.001</b> )
Anxiety	0.14 (0.27)	-0.29 (0.02)	0.07 (0.60)	0.19 (0.13)	0.29 (0.02)	-0.09 (0.48)	0.38 ( <b>&lt;0.01</b> )
Depression	0.32 ( <b>&lt;0.01</b> )	-0.29 (0.02)	0.004 (0.97)	0.31 (0.01)	0.40 (0.001)	-0.34 ( <b>&lt;0.01</b> )	0.42 ( <b>&lt;0.001</b> )
Social Isolation	0.13 (0.29)	-0.32 (0.01)	-0.06 (0.65)	-0.02 (0.86)	0.15 (0.22)	-0.17 (0.18)	0.31 ( <b>&lt;0.01</b> )
Peer Relationships	-0.07 (0.55)	0.21 (0.09)	0.45 ( <b>&lt;0.001</b> )	-0.03 (0.84)	0.04 (0.78)	0.10 (0.45)	-0.04 (0.76)

<sup>1</sup>Higher PROMIS® domain scores indicated worse severity for depression, anxiety, fatigue, and physical stress, but better mobility, self-efficacy, and peer relationships; <sup>2</sup>Higher B-IPQ scores indicate worse perceptions of SCD-related consequences, concerns, and emotional response, but better perceptions of understanding of SCD and personal control; <sup>3</sup>P-value ≤0.05 was statistically significant (highlighted in bold)

**Table 2: Brief-IPQ and Adherence Measures Categorical**

	VAS			ARMS			PMAS		
	Low (n = 29)	High (n = 38)	P value	Low (n = 38)	High (n = 29)	P value	Low (n = 33)	High (n = 34)	P value
B-IPQ Domains, median (IQR)									
Consequences	3 (2,4)	2 (1,4)	0.18	3 (2,4)	2 (1,3)	<b>&lt;0.01</b>	3 (2,4)	2 (1,3)	0.10
Personal Control	3 (2,4)	3 (3,4)	0.10	3 (3,4)	4 (3,4)	0.24	3 (2,4)	3 (3,4)	0.11
Treatment Control	4 (3,4)	5 (4,5)	<b>&lt;0.001</b>	4 (3,5)	5 (4,5)	0.03	4 (3,4)	5 (4,5)	<b>&lt;0.001</b>
Identity	3 (3,3)	2 (1,3)	0.03	3 (3,3)	2 (1,2)	<b>&lt;0.01</b>	3 (2,3)	2 (1,3)	0.09
Concerns	3 (2,4)	2.5 (1,3)	0.20	3 (2,4)	2 (2,3)	0.55	3 (2,4)	2 (2,3)	0.22
Understanding	4 (3,5)	4 (3,5)	0.44	4 (3,5)	4 (3,5)	0.66	4 (3,5)	4 (3,5)	0.19
Emotional Response	3 (1,4)	2 (0,3)	0.08	3(1,4)	1 (0,3)	0.04	2 (1,3)	2 (0,3)	0.56

PMAS: Low Adherence (< 38) and High Adherence: (≥38). ARMS: Low Adherence (≥12) and High Adherence: (<12). VAS: Low Adherence (<80) and High Adherence (≥80).

**Figure 1**

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