



The 65th ASH Annual Meeting Abstracts

POSTER ABSTRACTS

114. SICKLE CELL DISEASE, SICKLE CELL TRAIT AND OTHER HEMOGLOBINOPATHIES, EXCLUDING THALASSEMIAS: CLINICAL AND EPIDEMIOLOGICAL**Sickle Cell Health Awareness, Perspectives, and Experiences (SHAPE) Survey: Findings on the Burden of Sickle Cell Disease and Impact on the Quality of Life of Patients and Caregivers in Gulf Cooperation Council Countries**

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Introduction: Sickle cell disease (SCD) is an inherited disorder that adversely impacts the physical and emotional well-being of patients and caregivers, yet research on the impact of SCD on quality of life (QoL) is limited. In a systematic review the prevalence of SCD in Gulf Cooperation Council (GCC) countries ranged 0.24-5.8% (Abu-Shaheen et al. Expert Rev Hematol 2022). The 3 countries with the highest prevalence were Bahrain (1.2-2.1%, 2 studies), Saudi Arabia (0.24-4.5%, 15 studies), and Oman (5.8%, 1 study). The Sickle Cell Health Awareness, Perspectives, and Experiences (SHAPE) survey aimed to improve our understanding of the global burden of SCD. This analysis reports the experiences of patients with SCD and caregivers in GCC countries who participated in the SHAPE survey within the context of insights obtained globally.

Methods: The SHAPE survey included patients and caregivers from Brazil, Canada, France, Germany, the UK, the US, and 4 countries in the GCC (Bahrain, Oman [patients only], Saudi Arabia, and the United Arab Emirates [UAE]). Participants completed a 12-minute face-to-face survey (GCC only) or online survey (other countries) comprising close-ended questions about their experiences with SCD. Patients with SCD aged ≥ 12 years and caregivers aged ≥ 18 years supporting a patient with SCD were eligible.

Results: Of 919 patients and 207 caregivers globally who responded, 150 patients (16.3%; Saudi Arabia n=70, the UAE n=46, Bahrain n=19, and Oman n=15) and 50 caregivers (24.2%; Saudi Arabia n=25, the UAE n=17, and Bahrain n=8) were from GCC countries (Table). Symptoms experienced most frequently by patients in GCC countries and all surveyed patients, respectively, were fatigue/tiredness (91% vs 84%), bone aches (91% vs 69%), and vaso-occlusive crisis (VOC) pain (81% vs 71%). Compared with all surveyed patients, a significantly greater proportion of patients in GCC countries experienced fatigue/tiredness, bone aches, VOC pain, generalized pain, and signs of organ damage (Figure). Symptoms reported by a significantly smaller proportion of patients in GCC countries than by all surveyed patients were poor sleep/insomnia, poor appetite, memory/concentration issues, nausea, yellow eyes/nails/skin, vision difficulties/retinopathy, and leg ulcers. The top 3 symptoms patients in GCC countries reported as being most impactful in terms of the following categories were: (i) relationships/potential relationships (25% VOC pain, 19% low mood/feeling depressed, 16% bone aches); (ii) family (30% VOC pain, 12% bone aches, 11% low mood/feeling depressed); and (iii) long-term health prospects (29% bone aches, 23% VOC pain, 21% signs of organ damage). The average number of missed school/work days in the past month for patients was 3.9 in GCC

countries and 7.5 globally. A smaller proportion of patients in GCC countries were optimistic about their future compared with all surveyed patients (35% vs 52%).

The biggest concerns of caregivers of individuals with SCD were worsening of symptoms (54% in GCC countries vs 17% globally) and early loss of life (34% vs 39%). Approximately half of caregivers agreed that caring for a person with SCD had a big or noticeable impact on their family (52% in GCC countries vs 50% globally). The majority of caregivers agreed they were well-equipped to care for someone with SCD (80% in GCC countries vs 72% globally). A smaller proportion of caregivers in GCC countries were optimistic about the future of the person they care for with SCD compared with all surveyed caregivers (32% vs 62%).

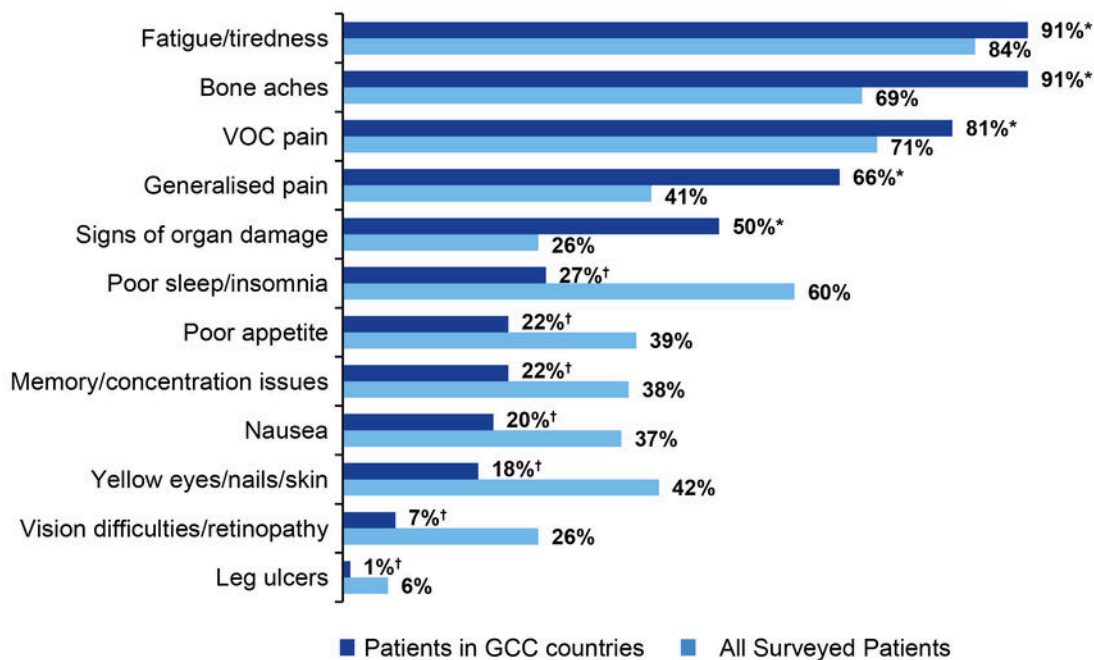
Conclusions: Findings from the SHAPE survey highlight the physical and emotional burden of SCD on patients and caregivers in GCC countries. Patients in GCC countries reported VOC pain, bone aches, and low mood/feeling depressed as symptoms that most affected their relationships/potential relationships and family. Caregivers in GCC countries expressed concerns about early loss of life and worsening SCD symptoms. Overall, these findings underline the need for effective treatments that alleviate symptoms and manage disease sequelae, and for additional resources to improve the QoL of patients and caregivers.

Disclosures Jastaniah: Novartis: Consultancy, Honoraria; Amgen: Consultancy, Honoraria; Bayer: Consultancy, Honoraria. **Hartfield:** Sickle Cell Disease Association of America, Inc.: Current Employment, Other: CEO and President. **James:** Sickle Cell Society: Current Employment, Other: CEO. **Tinga:** Sickle Cell Disease Association of Canada: Current Employment, Other: President. **Ingoli:** IST e.V.: Current Employment, Other: President. **de Montalembert:** Addmedica: Consultancy, Membership on an entity's Board of Directors or advisory committees; Novartis: Consultancy; Vertex: Consultancy, Other: Steering committee. **Costa:** Novartis: Consultancy. **Anderson:** Vertex: Consultancy; Pfizer: Consultancy, Research Funding, Speakers Bureau; Novo Nordisk: Consultancy; Novartis Pharmaceuticals Corporation: Consultancy. **Kunz:** Pfizer Inc.: Consultancy; Novartis: Consultancy; Bluebird Bio: Consultancy. **Odame:** Novo Nordisk: Consultancy; Novartis: Consultancy; Pfizer Inc.: Honoraria. **Lartey:** Ipsos Healthcare: Ended employment in the past 24 months. **Inusa:** Nova: Honoraria; Forma Therapeutics: Honoraria; Pfizer, Inc.: Honoraria, Other: Educational funding; AstraZeneca: Other: Educational funding; Bluebird Bio: Other: Educational funding; Celgene: Other: Educational funding; Novartis: Honoraria, Other: Educational funding; Cycleron: Honoraria; Novo Nordisk: Honoraria; Agios: Honoraria.

	Patients		Caregivers	
	GCC (n=150)	Total (n=919)	GCC (n=50)	Total (n=207)
Mean age, y	30.3	32.3	40.0	39.1
12 to <18, n (%)	0	64 (7)	0	0
18 to 39, n (%)	124 (83)	648 (71)	23 (46)	111 (54)
≥40, n (%)	26 (17)	207 (23)	27 (54)	96 (46)
Female, n (%)	75 (50)	659 (72)	27 (54)	149 (72)
Current employment/education status, n (%)				
Working full-time	23 (15)	257 (28)	13 (26)	83 (40)
Working part-time or self-employed	30 (20)	208 (23)	16 (32)	60 (29)
Student	28 (19)	170 (18)	1 (2)	7 (3)
Retired	4 (3)	34 (4)	1 (2)	7 (3)
Unemployed/not working	65 (43)	274 (30)	19 (38)	54 (26)
Highest educational achievement, n (%)				
Primary school	10 (7)	100 (11)	2 (4)	8 (4)
Secondary school	64 (43)	222 (24)	13 (26)	61 (29)
Technical college/apprenticeship	15 (10)	168 (18)	6 (12)	40 (19)
University degree	54 (36)	299 (33)	20 (40)	62 (30)
Masters/PhD	4 (3)	110 (12)	6 (12)	29 (14)

GCC=Gulf Cooperation Council

Figure. Symptoms Experienced by a Significantly Higher or Lower Proportion of Patients in GCC Countries Compared with All Surveyed Patients



* Indicates a significantly higher percentage of patients in GCC countries compared with all surveyed patients. † indicates a significantly lower percentage of patients in GCC countries compared with all surveyed patients. Statistical testing was based on a *t*-test with an α level of 0.05. Patients' responses to the question, "In the past year, which of the following signs and symptoms, if any, have you experienced?" GCC=Gulf Cooperation Council; VOC=vaso-occlusive crisis

Figure 1

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