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UK Patient Perceptions on the Symptomatic and Emotional Burden of Vaso-Occlusive Crises Resulting From Sickle Cell Disease

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INTRODUCTION

- Sickle cell disease (SCD) is characterised by vaso-occlusive crises (VOCs), which impact patients' quality of life (QoL) and increase the risk of organ damage and early death[1].
- There are limited published data describing the outcome of VOCs despite the chronic damage and fatality risk following a single VOC.
- Understanding the burden of SCD on patients can inform optimal disease management and improve patients' QoL.

AIM & METHOD

- The International Sickle Cell World Assessment Survey (SWAY) was a cross-sectional survey of 2,145 SCD patients and 365 healthcare professionals (HCPs) across 16 countries.
- This is the largest global survey of SCD patients ever conducted providing real-world insight into the burden of SCD, beyond what has been reported in the literature, in addition to the elements of treatment and management that are most important to the patient.
- Here we present data from UK patients.
- VOCs were defined as 'severe pain crises' and 'severe' was patient defined.
- Analyses were performed to evaluate impact of VOC burden on emotional wellbeing. VOC burden was grouped into 0-1, 2-4, 5-10, and 11+ VOCs experienced in last 12 months.
- Impact statements on emotional burden and activities of daily living were asked using a 7-point Likert scale with high agreement and satisfaction scores = 5–7.

Figure 1. Participant Categories:

Patients aged 6-11 participated by a proxy (parent/guardian/caregiver)

Patients aged 12 and above had the option either to complete themselves or via a proxy

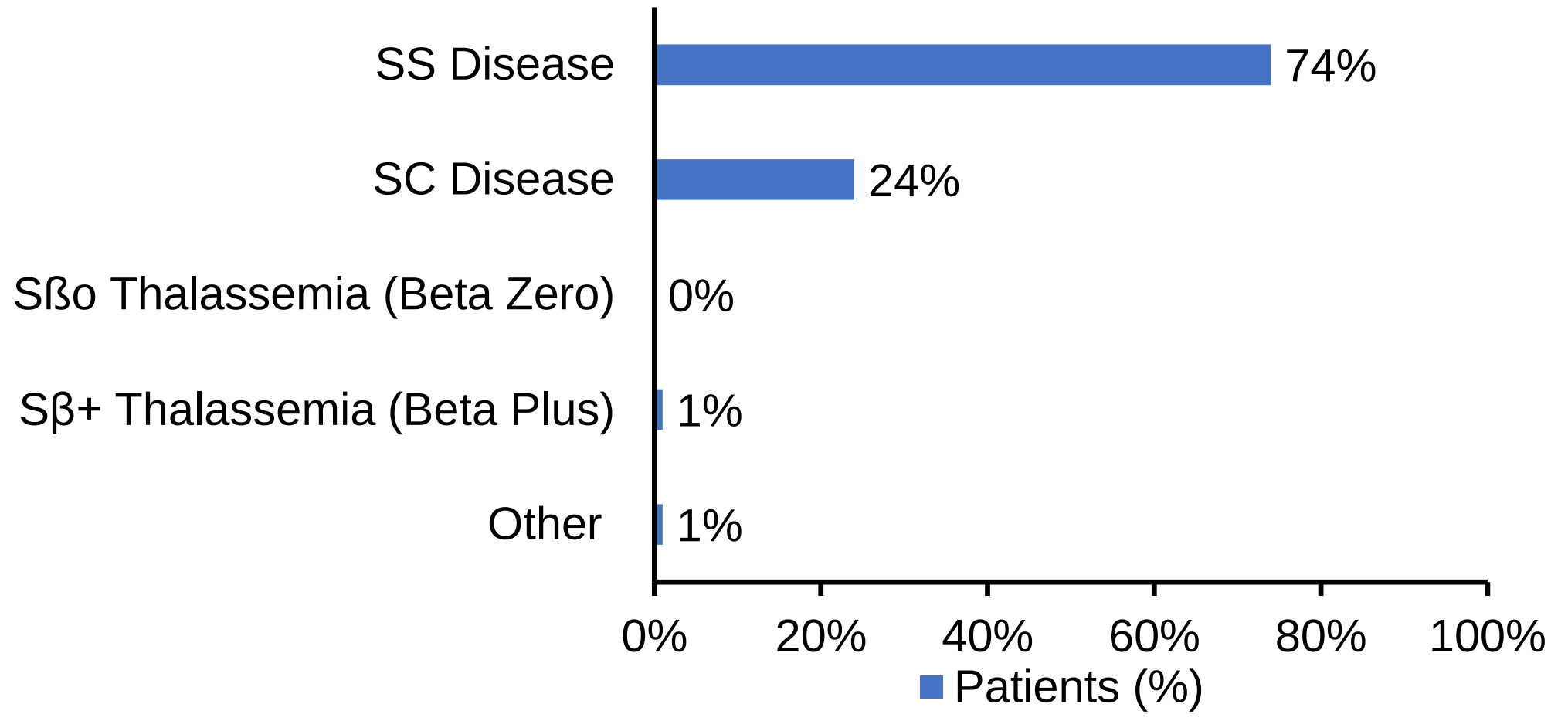
Treating healthcare professionals (HCPs)
• 57% Haematology specialists
• 43% General Practitioners

RESULTS

Demographics

- 299 UK patients completed the survey; 56% were female, mean (SD) age was 29.2 (12.4) years.
- Overall, 58 paediatric (<18) and 241 adult patients completed a survey.
- The mean (SD) age at diagnosis was 6.6 (7.98) years old, the majority of patients knew their genotype (91%), most having SS Disease (74%), as shown in ***Figure 2***.

Figure 2. SCD Genotype Diagnosed (n=272)

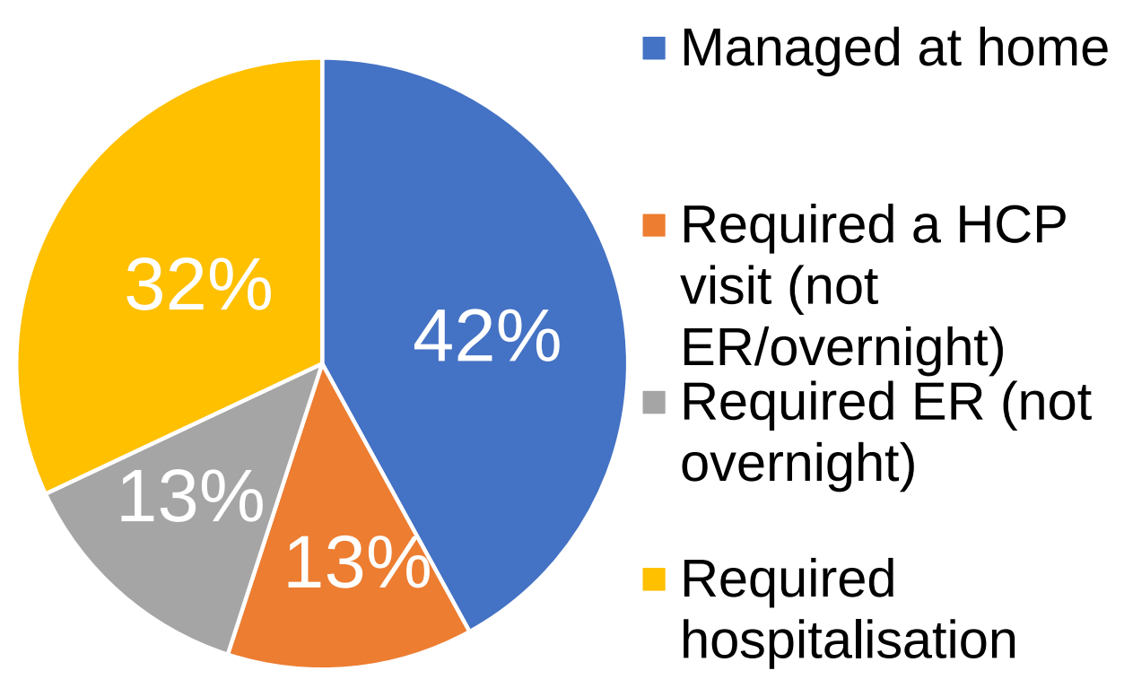


VOC Burden

- The majority (94%) of patients reported experiencing at least one VOC in the past 12 months, 40% experienced 5 or more, as shown in ***Figure 3***. Patients experienced multiple VOCs per year (mean 5.4, SD 5.1), highlighting a considerable VOC burden.
- For most episodes of VOC, health care support was required (42% of VOCs were managed at home), a third (32%) required hospitalisation, a considerable burden on health care resource utilisation (HCRU).

Figure 3. Incidence and Management of VOCs in Last 12 Months (n=299)

Number of VOCs in 12 Months	Patients, %
0	6
1	12
2-4	41
5-10	24
11+	16

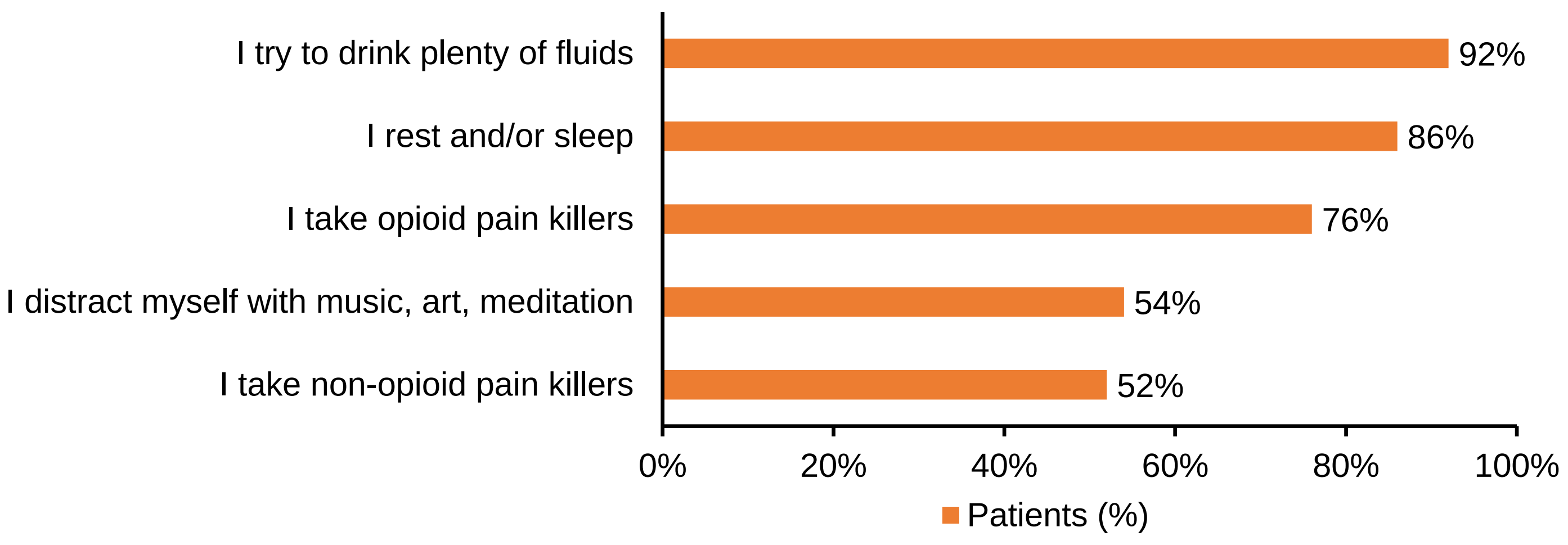


ER, emergency room; HCP, health care professional

Home Management of VOCs

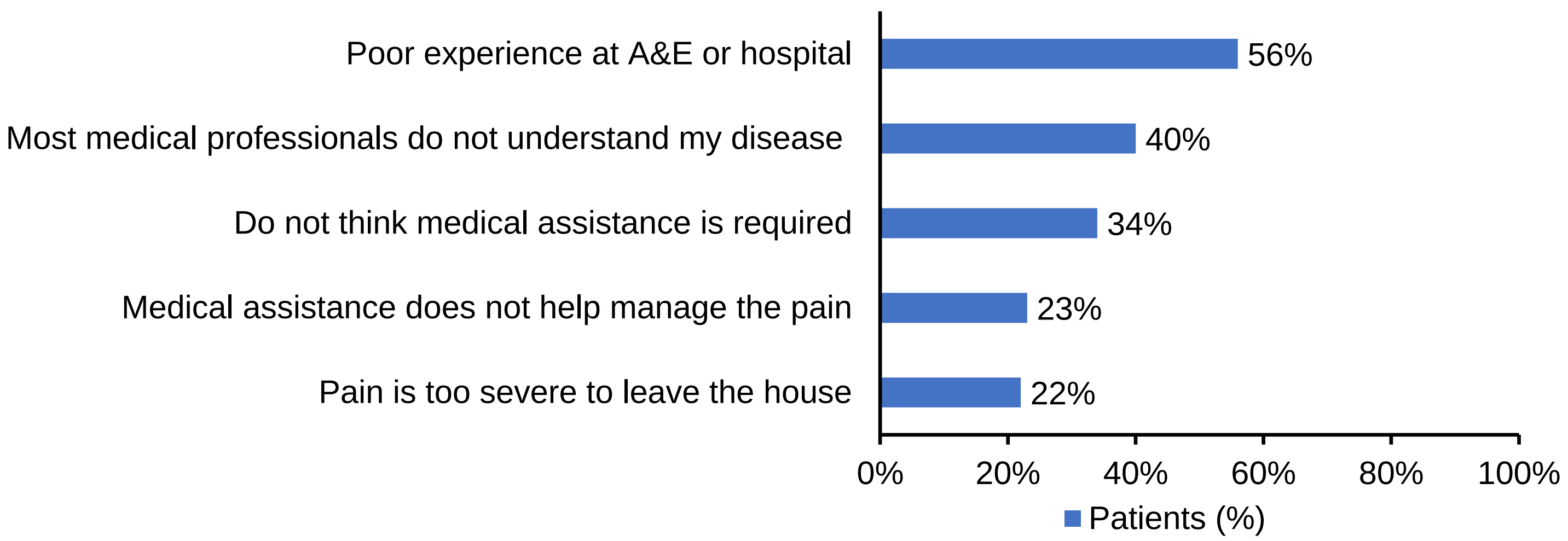
- ***Figure 4*** displays how patients manage VOCs at home (n=167), they do it mainly by drinking fluids (92%), resting/sleeping (86%), and taking opioid pain killers (76%).

Figure 4. Top 5 Ways to Manage VOCs at Home (n=167)



- The main motivations for managing VOCs at home were 'poor experience at A&E or hospital' (56%) and thinking 'medical professionals do not understand my disease' (40%), as shown in ***Figure 5***.

Figure 5. Top 5 Motivations for Managing VOCs at Home (n=167)



Emotional Well-being

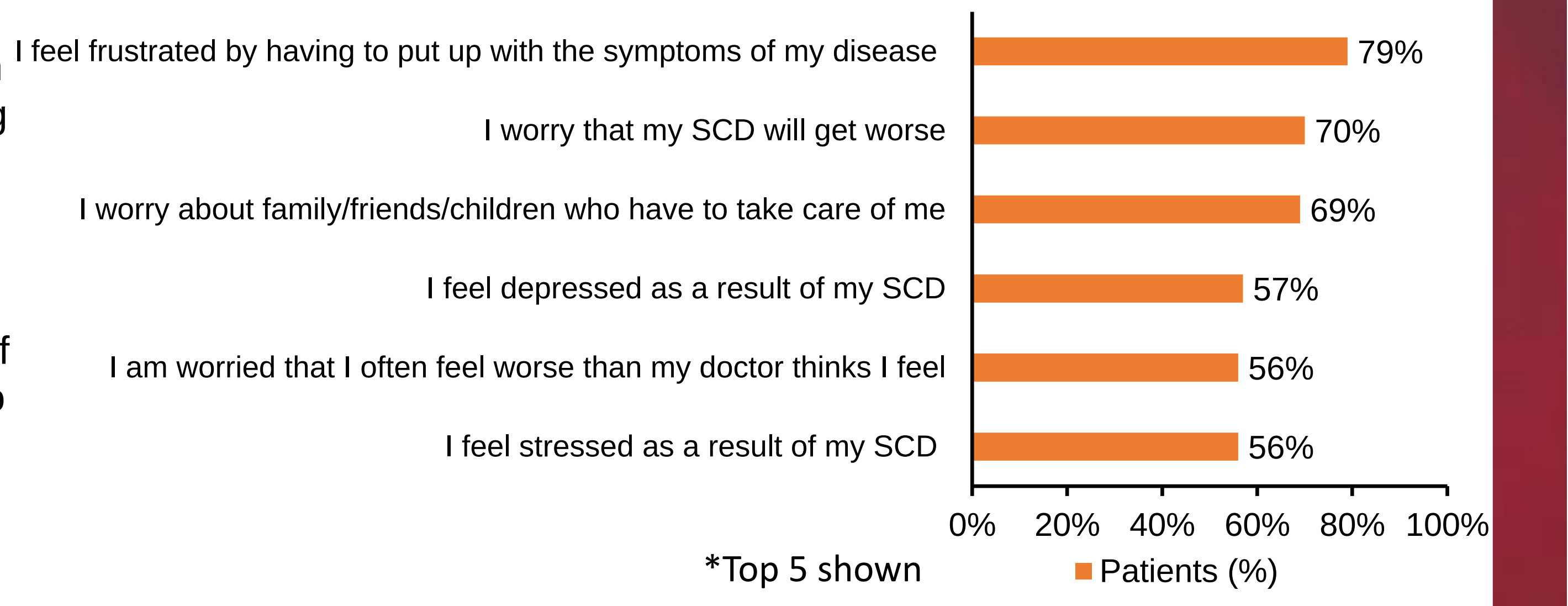
- The survey assessed the emotional burden of SCD for patients. Over two thirds of patients (69%) reported a high impact on emotional well-being, when asked "Overall, how does your SCD impact your emotional well-being?"
- Patients reported a higher emotional impact with increasing VOC burden, as shown in ***Figure 6***.

Figure 6. Impact of VOC Burden on Emotional Well-being

Number of VOCs in 12 Months	Patients, % Reporting High Impact (5-7)
0-1	52%
2-4	66%
5-10	77%
11+	86%

- Individual statements were rated by patients on the 7-point scale with the percentage reporting high impact (score 5–7) for each statement shown in ***Figure 7***.
- Patients reported a higher impact in terms of frustration with having to put up with symptoms (79%) and worry that their SCD will get worse (70%).

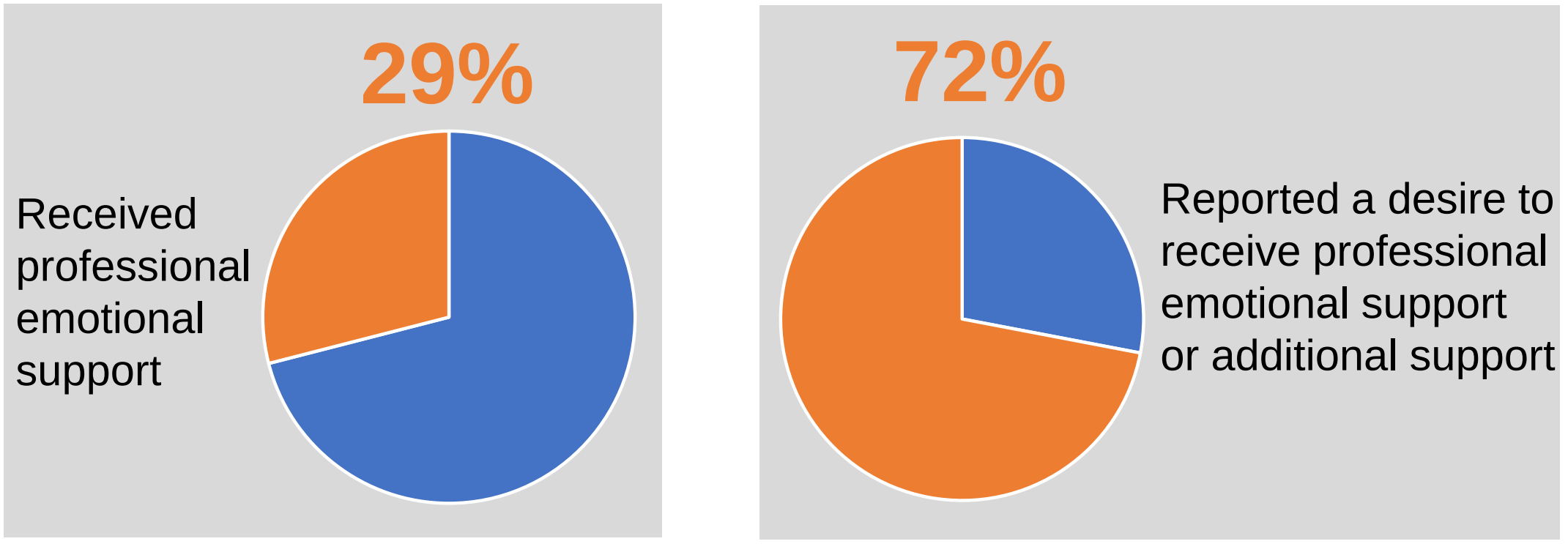
*Figure 7. Emotional Impact of SCD (n=299)**



*Top 5 shown

- Less than a third (29%) of all patients received professional emotional support (e.g. psychiatrist, psychologist, counselling), whilst 72% of all patients reported a desire to receive additional support, as shown in ***Figure 8***.

Figure 8. Professional Emotional Support Received (n=299)



Impact on Daily Life

- Patients reported SCD had a high impact on activities of daily living, including social and role functioning, as shown in ***Figure 9***.

Figure 9. Impact of SCD on Daily Life (n=299)

Daily Life Category	% of Patients Reporting High Impact (5-7)
Daily activities (i.e. food preparation, housework, gardening, taking care of children, oral hygiene)	43%
Family or social life (meeting friends/family for activities, hobbies)	47%
Relationship with spouse/partner*	39%
Relationship with other family members	32%
Sexual desire/activity*	37%

***N=241**

CONCLUSIONS

- This analysis indicates that SCD patients in the UK experience a high VOC symptom burden, a large proportion of which patients choose to manage at home due to fear of poor experience at A&E or hospital.
- There is indication that SCD has a tremendous impact on the emotional health of patients, which is currently not being addressed adequately, and remains an area for significant improvement.
- Timely and effective pain relief in A&E is vital for people living with SCD. It is critical that A&E departments follow the *NICE guidelines.

*National Institute for Health and Care Excellence

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1 **Rizio A et al**. Pro66 Impacts of Vaso-occlusive Crises on the Quality of Life and Healthcare Resource Utilisation of Patients with Sickle Cell Disease. *Value in Health 2019; 22;: S347-S348*

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