The aim of the International Sickle Cell World Assessment Survey (SWAY) was to assess the impact of SCD on the daily lives of patients. Patient surveys were either self-completed or completed by proxy (mandatory for patients aged 6–11 years, available for older patients if required). SWAY included 2145 SCD patients from 16 countries across five continents. 365 HCPs were included in the survey.

**SWAY patient region of residence**

<table>
<thead>
<tr>
<th>Region</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>North America</td>
<td>513</td>
</tr>
<tr>
<td>South America</td>
<td>260</td>
</tr>
<tr>
<td>Europe</td>
<td>557</td>
</tr>
<tr>
<td>Africa</td>
<td>519</td>
</tr>
<tr>
<td>Asia and the Middle East</td>
<td>296</td>
</tr>
</tbody>
</table>

Genotypes were self-reported* by patients in SWAY, including HbSS (n=1042 [48%]), HbSC (n=446 [21%]), HbSβ+ (n=60 [3%]), HbSβ0 (n=54 [3%]) and other/unknown (n=543 [25%]).

HCPs in SWAY included hematologists (41%), hematologist-oncologists (28%) and general practitioners (14%).

The average age (SD) of patients was 25 (13.1) years. Youngest age: 6 years old. Oldest age: 75 years old.

48% of patients were male. 52% of patients were female.

*Only reported if certain of genotype

---

The aim of the International Sickle Cell World Assessment Survey (SWAY) was to assess the impact of SCD on the daily lives of patients. Patient surveys were either self‐completed or completed by proxy (mandatory for patients aged 6–11 years, available for older patients if required). SWAY included 2145 SCD patients from 16 countries across five continents. 365 HCPs were included in the survey.

The average age (SD) of patients was 25 (13.1) years. 48% of patients were male and 52% were female.

Genotypes were self-reported by patients in SWAY, including HbSS (n=1042 [48%]), HbSC (n=446 [21%]), HbSβ0 (n=54 [3%]), HbSβ+ (n=60 [3%]), and other/unknown (n=543 [25%]).

Canada (n=120), USA (n=384), Panama (n=9), Brazil (n=260), France (n=76), Germany (n=94), Italy (n=55), Netherlands (n=33), UK (n=299), Ghana (n=255), Nigeria (n=264), Bahrain (n=50), India (n=123), Lebanon (n=39), Oman (n=52), Saudi Arabia (n=32), North America (n=513), South America (n=260), Europe (n=557), Africa (n=519), Asia and the Middle East (n=296).

*Only reported if certain of genotype
HCP, healthcare professional; SCD, sickle cell disease; SD, standard deviation.
The average age (SD) of patients was 25 (13.1) years. The aim of the International Sickle Cell World Assessment Survey (SWAY) was to assess the impact of SCD on the daily lives of patients. Patient surveys were either self-completed or completed by proxy (mandatory for patients aged 6–11 years, available for older patients if required). SWAY included 2145 SCD patients from 16 countries across five continents, 365 HCPs were included in the survey. HCPs in SWAY included hematologists (41%), hematologist-oncologists (28%) and general practitioners (14%). Genotypes were self-reported* by patients in SWAY, including HbSS (n=1042 [48%]), HbSC (n=446 [21%]), HbSβ+ (n=60 [3%]), HbSβ0 (n=54 [3%]) and other/unknown (n=543 [25%]).

48% of patients were male and 52% of patients were female.

SWAY was a multi-country, cross-sectional survey developed by international SCD experts, patient advocacy groups and Novartis.

“We know some things about sickle cell disease, we’ve discovered a lot in the past 30 years on pathophysiology and clinical manifestations, but we felt we only see the tip of the iceberg and a lot more needs to be understood.”

Raffaella Colombatti, MD, PhD
Pediatric Hematologist
Clinic of Pediatric Hematology
University of Padova
SWAY Steering Committee member

“Understanding what is common to people who live with sickle cell across the globe and how we might work together to address those issues is very important.”

John James, OBE
Chief Executive
Sickle Cell Society
SWAY Steering Committee member

*SOnly reported if certain of genotype
HCP, healthcare professional; SCD, sickle cell disease; SD, standard deviation
Patients in SWAY reported a mean (SD) of 5.3 (6.8) VOCs per year\textsuperscript{1,2}

- VOCs in SWAY were defined as severe pain crises and were self-reported\textsuperscript{1–3}
- Patients reported a median (IQR) of 3.0 (2.0, 6.0) VOCs per year\textsuperscript{3}

### Number of VOCs in 12 months (N=2142*)\textsuperscript{1,2}

<table>
<thead>
<tr>
<th>Number of VOCs</th>
<th>Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>9%</td>
</tr>
<tr>
<td>1</td>
<td>12%</td>
</tr>
<tr>
<td>2–4</td>
<td>39%</td>
</tr>
<tr>
<td>≥5</td>
<td>39%</td>
</tr>
</tbody>
</table>

### Where were VOCs managed? (N=2142*)\textsuperscript{1,2}

- 18% ER
- 33% Hospital (overnight stay)
- 24% Community\textsuperscript{†}
- 24% Home

### Most common reasons for home management (N=881)

<table>
<thead>
<tr>
<th>Reason</th>
<th>Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Previous poor hospital or ER experience</td>
<td>39%</td>
</tr>
<tr>
<td>Patient thought medical assistance was not needed</td>
<td>30%</td>
</tr>
<tr>
<td>Patient thought medical professionals do not understand SCD</td>
<td>26%</td>
</tr>
<tr>
<td>Cost of hospital treatment</td>
<td>20%</td>
</tr>
<tr>
<td>Pain too severe to leave home</td>
<td>19%</td>
</tr>
</tbody>
</table>

### Common strategies employed by patients (N=881) for the management of VOCs at home

- Rest/sleep: 74%
- Drinking fluids: 74%
- Opioid-based analgesia: 58%
- Warm baths/showers: 48%

\textsuperscript{*VOC data not available for three patients; †Community includes assistance from a pharmacist, general practitioner or specialist, but does not include ER treatment or overnight hospitalization; ER, emergency room; IQR, interquartile range; VOCs, vaso-occlusive crises
Patients in SWAY reported a mean (SD) of 5.3 (6.8) VOCs per year\(^1,2\)

- VOCs in SWAY were defined as severe pain crises and were self-reported\(^1-3\)
- Patients reported a median (IQR) of 3.0 (2.0, 6.0) VOCs per year\(^3\)

<table>
<thead>
<tr>
<th>Number of VOCs in 12 months (N=2142*)(^1,2)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
</tr>
<tr>
<td>0</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Where were VOCs managed? (N=2142*)(^1,2)</th>
</tr>
</thead>
<tbody>
<tr>
<td>33%</td>
</tr>
<tr>
<td>18%</td>
</tr>
<tr>
<td>12%</td>
</tr>
<tr>
<td>24%</td>
</tr>
<tr>
<td>5%</td>
</tr>
</tbody>
</table>

- The SWAY survey showed that there was geographic variability in the incidence of VOCs\(^3\)
- The countries that reported the highest number of VOCs/patient (median [IQR]) were:
  - Lebanon (12.0 [10.0, 15.0])
  - USA (6.0 [3.0, 10.0])
  - Bahrain (5.0 [3.0, 40.0])

*VOC data not available for three patients; \(^1\)Community includes assistance from a pharmacist, general practitioner or specialist, but does not include ER treatment or overnight hospitalization; ER, emergency room; IQR, interquartile range; VOCs, vaso-occlusive crises
Patients in SWAY reported a mean (SD) of 5.3 (6.8) VOCs per year\(^1,2\)

- VOCs in SWAY were defined as severe pain crises and were self-reported\(^1\)–\(^3\)
- Patients reported a median (IQR) of 3.0 (2.0, 6.0) VOCs per year\(^3\)

**Number of VOCs in 12 months (N=2142*)\(^1,2\)**

![Graph showing number of VOCs managed by country: Italy (50%), Canada (44%), UK (42%).]

**Home management of VOCs by country\(^3\)**

- Countries that reported the highest proportion of VOCs managed at home:
  - Italy (50%)
  - Canada (44%)
  - UK (42%)

- Main reason for managing VOCs at home:
  - Thinking medical assistance was not required
  - Poor experience at the ER or hospital

**Common strategies employed by patients (N=881) for the management of VOCs at home**

- Rest/sleep: 74%
- Drinking fluids: 74%
- Opioid-based analgesia: 58%
- Warm baths/showers: 48%

\(^*\)VOC data not available for three patients; \(^*\)Community includes assistance from a pharmacist, general practitioner or specialist, but does not include ER treatment or overnight hospitalization; \(^*\)ER, emergency room; IQR, interquartile range; VOCs, vaso-occlusive crises

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Number of VOCs in 12 months (N=2142*)\(^1,2\)

Where were VOCs managed? (N=2142*)\(^1,2\)

Home management of VOCs by age\(^3\)

- Patients aged ≤25 years
  - Main reason for managing VOCs at home: Thinking medical assistance was not required
  - Most common way to manage VOCs at home: Resting and/or sleeping

- Patients aged ≥26 years
  - Main reason for managing VOCs at home: Poor experience at the ER or hospital
  - Most common way to manage VOCs at home: Trying to drink plenty of fluids

Common strategies employed by patients (N=881) for the management of VOCs at home:

- Rest/sleep: 74%
- Drinking fluids: 74%
- Opioid-based analgesia: 58%
- Warm baths/showers: 48%

*VOC data not available for three patients; \(^*\)Community includes assistance from a pharmacist, general practitioner or specialist, but does not include ER treatment or overnight hospitalization; ER, emergency room; IQR, interquartile range; VOCs, vaso-occlusive crises

Patients in SWAY reported a mean (SD) of 5.3 (6.8) VOCs per year\textsuperscript{1,2}

There’s a lot more VOCs happening than we’ve been able to capture from the typical way we look at VOCs…we’ve found that patients don’t always go to the hospital and handle a lot of the pain on their own

Ify Osunkwo, MD, MPH
Principal Investigator, Patient-Centred Outcomes Research Institute (PCORI); Medical Director, Sickle Cell Enterprise, Levine Cancer Institute, Carolina Healthcare System; SWAY Steering Committee chair

Most common reasons for home management (N=881)

- Previous poor hospital or ER experience
- Patient thought medical assistance was not needed
- Patient thought medical professionals do not understand SCD
- Cost of hospital treatment
- Pain too severe to leave home

Common strategies employed by patients (N=881) for the management of VOCs at home

- Rest/sleep 74%  
- Drinking fluids 74%  
- Opioid-based analgesia 58%  
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\textsuperscript{1}VOC data not available for three patients; \textsuperscript{2}Community includes assistance from a pharmacist, general practitioner or specialist, but does not include ER treatment or overnight hospitalization; ER, emergency room; IQR, interquartile range; VOCs, vaso-occlusive crises

Patients in SWAY reported a mean (SD) of 5.3 (6.8) VOCs per year\textsuperscript{1,2}

- VOCs in SWAY were defined as severe pain crises and were self-reported\textsuperscript{1–3}
- Patients reported a median (IQR) of 3.0 (2.0, 6.0) VOCs per year\textsuperscript{3}

<table>
<thead>
<tr>
<th>Number of VOCs in 12 months (N=2142*)\textsuperscript{1,2}</th>
<th>Where were VOCs managed? (N=2142*)\textsuperscript{1,2}</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients (%)</td>
<td>18% ER</td>
</tr>
<tr>
<td>0–4</td>
<td>33% Hospital (overnight stay)</td>
</tr>
<tr>
<td>5–9</td>
<td>24% Community†</td>
</tr>
<tr>
<td>10–15</td>
<td>24% Home</td>
</tr>
<tr>
<td>16–20</td>
<td>9%</td>
</tr>
<tr>
<td>21–25</td>
<td>12%</td>
</tr>
<tr>
<td>26–30</td>
<td>39%</td>
</tr>
<tr>
<td>31–40</td>
<td>39%</td>
</tr>
<tr>
<td>41–45</td>
<td></td>
</tr>
<tr>
<td>46–50</td>
<td></td>
</tr>
<tr>
<td>51–55</td>
<td></td>
</tr>
<tr>
<td>56–60</td>
<td></td>
</tr>
<tr>
<td>61–65</td>
<td></td>
</tr>
<tr>
<td>66–70</td>
<td></td>
</tr>
<tr>
<td>71–75</td>
<td></td>
</tr>
<tr>
<td>76–80</td>
<td></td>
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<tr>
<td>81–85</td>
<td></td>
</tr>
<tr>
<td>86–90</td>
<td></td>
</tr>
<tr>
<td>91–95</td>
<td></td>
</tr>
<tr>
<td>96–100</td>
<td></td>
</tr>
</tbody>
</table>

VOCs in SWAY were defined as severe pain crises and were self-reported. Patients reported a median (IQR) of 3.0 (2.0, 6.0) VOCs per year.

For one reason or another, either due to a lack of treatment options or a lack of quality interaction with providers in general, (patients) are choosing to stay at home and suffer.

\textsuperscript{*VOC data not available for three patients; †Community includes assistance from a pharmacist, general practitioner or specialist, but does not include ER treatment or overnight hospitalization; ER, emergency room; IQR, interquartile range; VOCs, vaso-occlusive crises


Biree Andemariam, MD
Director, New England Sickle Cell Institute of Connecticut Health Center; Chief Medical Officer, Sickle Cell Disease Association of America; SWAY Steering Committee member
The most common ongoing treatment was **folic acid** and the top treatment goal was **improving quality of life**.

**Common ongoing treatments reported by SCD patients (N=2123)**

- Folic acid: 59%
- Over-the-counter pain medication: 37%
- Anti-inflammatories: 34%
- Vitamin D: 34%
- Opioids: 34%
- Antibiotics: 33%
- Hydroxyurea: 31%

**Common treatment goals for patients with SCD (N=2145)* and HCPs (N=365)**

- Improving quality of life: 55% of patients, 64% of HCPs
- Prevent worsening of SCD: 43% of patients, 32% of HCPs
- Reducing number of VOCs: 30% of patients, 43% of HCPs

**Opioid use in SWAY**

<table>
<thead>
<tr>
<th>Type of opioid use (number of patients)</th>
<th>North America (n=513)</th>
<th>South America (n=260)</th>
<th>Europe (n=557)</th>
<th>Middle East (n=173)</th>
<th>Africa (N=519)</th>
<th>Asia (n=123)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ever</td>
<td>76%</td>
<td>37%</td>
<td>70%</td>
<td>62%</td>
<td>26%</td>
<td>8%</td>
</tr>
<tr>
<td>Current</td>
<td>58%</td>
<td>20%</td>
<td>45%</td>
<td>38%</td>
<td>11%</td>
<td>2%</td>
</tr>
<tr>
<td>Patients reporting chronic pain‡</td>
<td>34%</td>
<td>28%</td>
<td>34%</td>
<td>18%</td>
<td>2%</td>
<td>7%</td>
</tr>
<tr>
<td>Patients reporting poor health‡</td>
<td>19%</td>
<td>22%</td>
<td>15%</td>
<td>10%</td>
<td>16%</td>
<td>13%</td>
</tr>
</tbody>
</table>

**Higher use of opioids in North America, the Middle East and Europe than in South America, Africa and Asia corresponded to a greater degree of chronic pain but not reduced reported health status**

*Patients asked to select their “top three” treatment goals; ‡≥5 days of background pain in a week; †Score of 1–3 on a 7-point likert scale

Although patients were **satisfied with their doctor’s management of their SCD**, the majority of patients and HCPs have **treatment concerns**

- SCD specialists were the main type of HCP responsible for management of patients (60%)\(^1\)
- 76% of patients were satisfied with the frequency of their interactions with their doctor and 70% were confident in proper assessment/treatment by their doctor*\(^1\)

- Only **49%** of HCPs (N=365) reported high satisfaction with treatment options*\(^2\)
- HCPs reported that the main reason for treatment dissatisfaction was limited treatment options (90% of respondents)

- **66%** of patients reported high satisfaction with their treatment*\(^2\)
- 72% of patients agreed with the statement ‘I wish there was an alternative treatment to my current pain medication’*\(^2\)

### Patients’ dissatisfaction with treatments*\(^1\)

<table>
<thead>
<tr>
<th>Percentage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;50%</td>
<td>of patients substantially worry about the <strong>short-term</strong> effects of their treatment (N=2123)</td>
</tr>
<tr>
<td>&gt;60%</td>
<td>of patients substantially worry about the <strong>long-term</strong> effects of their treatment (N=2123)</td>
</tr>
<tr>
<td>62%</td>
<td>of opioid-receiving patients substantially worry about how they are perceived (N=919)</td>
</tr>
</tbody>
</table>

*Percentage of patients/HCPs reporting high impact (score 5–7) on a 7-point likert scale
Patients experienced a number of **symptoms** and **complications**, with SCD also having a **high impact on the patient’s daily life**

### Most commonly experienced symptoms and complications†

In SWAY, fatigue/tiredness was the most commonly reported symptom in the past month and fever was the most commonly reported complication ever experienced (VOCs were assessed separately)*

#### Symptoms experienced in past month
- **65%** Fatigue/tiredness
- **51%** Bone aches
- **47%** Headaches

#### Complications ever experienced
- **64%** Fever
- **56%** Joint issues
- **54%** Infection

Patients reported that background pain was present on average 2.8 days per week (SD=2.2)

### Impact of SCD on daily life (percentage of patients reporting high impact, †N=2145)†

<table>
<thead>
<tr>
<th>Family/social life</th>
<th>Sexual desire/activity‡</th>
<th>Household daily activities</th>
<th>Relationships with spouse/partner‡</th>
<th>Avoidance of intense activity</th>
<th>Avoidance of mild activity</th>
</tr>
</thead>
<tbody>
<tr>
<td>(41%)</td>
<td>(31%)†</td>
<td>(38%)</td>
<td>(32%)</td>
<td>(61%)</td>
<td>(26%)</td>
</tr>
</tbody>
</table>

### Patients reported that SCD had an impact on their work (N=968)†
- **53%** reduced their hours
- **57%** seriously considered reducing their hours at work
- **43%** considering terminating employment

### Patients reported a high impact of SCD on school (N=1376)‡†
- **51%** said SCD impacted their achievement
- **46%** said SCD reduced attendance at school§
- **41%** said SCD decreased their motivation at school

---

*Symptoms and complications included in the survey were selected by the steering committee; †Percentage of patients reporting high impact (score 5–7) on a 7-point likert scale; ‡N=1376 (patients ≥18 years); §N=769 (patients <18 years)

1. Osunkwo I et al. Poster 2297 presented at ASH 2019
Patients experienced a number of symptoms and complications, with SCD also having a high impact on the patient’s daily life.
## SCD has a substantial impact on emotional wellbeing

### Patient and HCP perceptions of the emotional impact of SCD*1,2

<table>
<thead>
<tr>
<th>Perception</th>
<th>Patient perception (N=2145)</th>
<th>HCP perception (N=365)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Overall, SCD impacts emotional wellbeing</td>
<td>60%</td>
<td>91%</td>
</tr>
<tr>
<td>Feel frustrated by having to put up with the symptoms of SCD</td>
<td>58%</td>
<td>82%</td>
</tr>
<tr>
<td>Worry that their SCD will get worse</td>
<td>58%</td>
<td>79%</td>
</tr>
<tr>
<td>Feel stressed as a result of SCD</td>
<td>49%</td>
<td>76%</td>
</tr>
<tr>
<td>Feel depressed as a result of SCD</td>
<td>44%</td>
<td>74%</td>
</tr>
<tr>
<td>Feel helpless as a result of SCD</td>
<td>43%</td>
<td>71%</td>
</tr>
<tr>
<td>Worry about dying as a result of SCD</td>
<td>48%</td>
<td>70%</td>
</tr>
<tr>
<td>Feel anxious/nervous as a result of SCD</td>
<td>44%</td>
<td>70%</td>
</tr>
</tbody>
</table>

### A greater proportion of HCPs reported high emotional impact compared with patients

- Patients may perceive a reduced level of emotional and physical wellbeing as normal, considering SCD is present from birth
- There may be a lack of discussion between patients and HCPs
- There is potential bias caused by HCPs being more likely to see patients on days when they are unwell

- Of patients (N=2145) received professional emotional support (eg psychiatrist, psychologist, counseling)1: 36%
- Of patients (N=2145) reported a desire to receive professional emotional support or additional support1: 62%

*Individual statements were rated by patients on a 7-point scale with the percentage reporting high impact (score 5–7) for each statement. HCPs were asked to rate the same statements from the viewpoint of their patients. 1. Osunkwo I et al. Poster 2297 presented at ASH 2019; 2. Trimnell C et al. Abstract EP1539 accepted by EHA 2020
The SWAY survey demonstrates the substantial global impact of SCD on patients’ daily lives.

The SWAY survey also provides evidence of geographic variability in the incidence and management of VOCs, with nearly a quarter of patients managing VOCs at home.

Patients and HCPs have treatment concerns, indicating a need for additional and alternative treatments.

SCD has a considerable impact on patients daily lives and emotional wellbeing.