The Impact of Living with Sickle Cell Disease in the UK

What is sickle cell disease (SCD)?

SCD is a complex, genetic blood disorder which is inherited from both parents.[1]

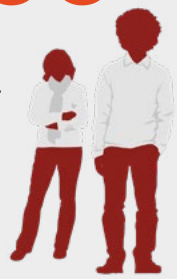
The main symptoms of sickle cell are anaemia, increased risk of serious infections, fatigue and episodes of severe pain known as sickle cell crisis or vaso-occlusive crisis.[2] VOCs occur due to increased interactions between different types of cells, including sickled red blood cells, other blood cells and the cells that line the blood vessel wall. These interactions cause painful blockages in the blood vessels.[3]

Who is affected by SCD?

APPROXIMATELY

15,000

people in the **UK have SCD**[4]



APPROXIMATELY

270

babies with **SCD are born** in the UK every year[4]

SCD

mainly **affects people of African or African-Caribbean origin in the UK.** However, the sickle gene is found in all ethnic groups[5]

About the International Sickle Cell World Assessment Survey (SWAY)

Cross-sectional survey of...

- **2,145** SCD patients
- **365** healthcare professionals (HCPs)
- **16** countries...

...assessing the **impact of sickle cell disease** on the daily life of patients, including physical symptoms, emotional wellbeing and economic burden.

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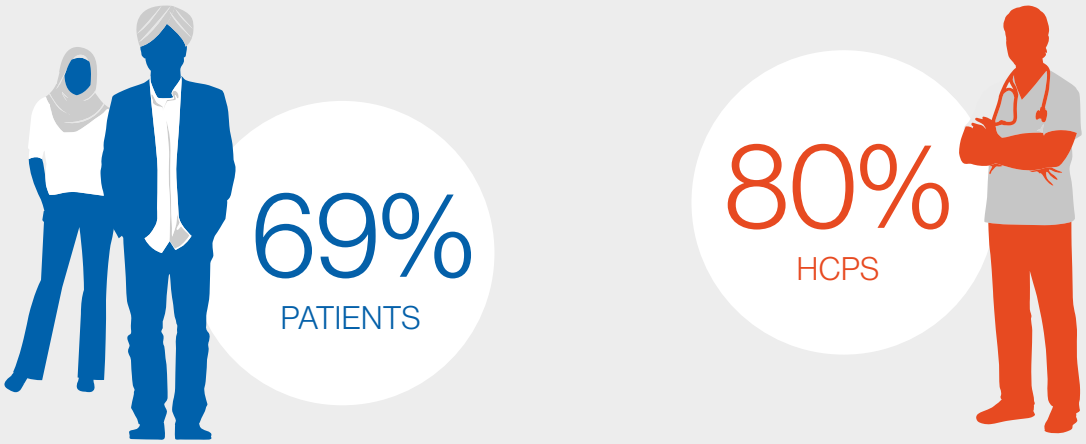
UK SWAY findings

Data from **299 UK patients**, who were recruited via treating HCPs and The Sickle Cell Society patient charity.

Quality of life

Patients were asked, other than a cure for sickle cell, **what are your 3 most important treatment goals?**
The most cited treatment goal for both patients and HCPs was...

'Improving quality of life'[6]



Comparative importance of additional treatment goals.[6] Patients vs HCPs*

	PATIENTS	HCPs
1. Improving quality of life	69%	80%
2. Improvement in overall symptoms	34%	37%
3. Prevent worsening of SCD	57%	10%
4. Reduce chronic pain levels	12%	40%
5. Reduce VOCs	21%	37%

Physical burden of SCD

High sickle cell crisis (VOC) burden

69% of patients reported high impact on **emotional wellbeing**[7]

94% of patients reported at least **one** VOC in the **past 12 months**[7]

Of all VOC crisis, **32%** lead to **hospitalisation**[7]

42% of VOCs are **managed at home**[7]

Reasons why people manage VOCs at home

- Belief that medical professionals do not understand sickle cell (40%)[7]
- Poor prior experiences at hospitals (56%)[7]

Complications of SCD

SCD increases an individuals risk of...

- Stroke[1]
- Acute chest syndrome[1]
- Blindness[1]
- Bone damage[1]
- Priapism[1]

SCD can also lead to organ damage, including the heart, lungs, kidneys, liver and spleen[1]

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Sickle cell impacts productivity at work

Patients reported that their disease has a high impact on their ability to work...

74% seriously considered **reducing their hours at work**...[8]

...**with** 61% having done so[8]

61% have considered **terminating their job**[8]

58% reported **reduced attendance** at work as a result of their sickle cell[8]

72% of patients reported their disease has **limited them to certain careers**[8]

"The SWAY study reveals the substantial burden UK sickle cell patients carry, and highlights where management of symptoms can be improved, learnings shared, and best practices established. This research is a vital tool in optimising both communication and care, and offers the chance to collectively improve management of the disease, improve health outcomes, and ultimately people's quality of life."

John James OBE, *Chief Executive, The Sickle Cell Society*

References: 1. Sickle Cell Society. About Sickle Cell Disease. Available from https://www.sicklecellsociety.org/about-sickle-cell/. Date accessed: June 2020 **2.** National health service (2019). Symptoms – Sickle cell disease. Available at: https://www.nhs.uk/conditions/sickle-cell-disease/symptoms/. Date accessed: June 2020 **3.** Kaul DK, et al. (2009). Sickle red cell - Endothelium interactions. Microcirculation, 16, 97-111. **4.** Sickle Cell Society. World Sickle Cell Day 2019. Available from: https://www.sicklecellsociety.org/wscd19/. Date accessed: June 2020 **5.** National Institute for Health and Care Excellence. (2014). Sickle cell disease. **6.** Inusa B, et al. Patients' and Health Care Professionals' Perspectives on Treatment Goals in Sickle Cell Disease – A UK Perspective: Results From the International Sickle Cell World Assessment Survey (SWAY). Abstract published in the British Journal of Haematology. Abstract BSH2020-PO-037 **7.** Inusa B, et al. UK Patient perceptions on the symptomatic and emotional burden of vaso-occlusive crises resulting from Sickle cell disease. Abstract published in the British Journal of Haematology. Abstract BSH2020-PO-079A **8.** Inusa B, et al. UK Results from the International Sickle Cell World Assessment Survey (SWAY): Patients with Sickle Cell Disease (SCD) Experience Work Impairment and Decreased Productivity. Abstract published in the British Journal of Haematology. Abstract BSH2020-OR-031

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