Press Release
For general media release

UK Findings from SHAPE Survey Highlight Physical, Societal and Economic Inequalities for People Living with Sickle Cell Disease (SCD)

- *Survey data released one year after publication of the "No One's Listening Report" by the All-Party Parliamentary Group on Sickle Cell and Thalassaemia and the Sickle Cell Society.*
- *SCD restricts peoples' ability to participate in education and employment and impacts earning potential, overall wellbeing and mental health; patients miss the equivalent of over three months of work or school every year due to living with SCD.*

Walton Oaks, 16 November 2022 – One year on since publication of the ground-breaking No One's Listening Report on sickle cell care in the UK, full results from the **S**ickle Cell **H**ealth **A**wareness, **P**erspectives and **E**xperiences (SHAPE) survey[1] reveal that significant healthcare inequalities for people in the UK living with sickle cell disease (SCD) persist. Findings from the survey, which was developed by Global Blood Therapeutics (GBT), a wholly owned subsidiary of Pfizer, supports the conclusions of the 'No One's Listening Report' published in November 2021 by the All-Party Parliamentary Group (APPG) on Sickle Cell and Thalassaemia in partnership with the Sickle Cell Society, which highlighted "serious care failings" in the healthcare of people in the UK with SCD. The survey findings show that SCD significantly impacts many aspects of the lives of people living with the condition, including their overall physical and mental wellbeing and their ability to attend and be successful at school or work, therefore reducing their earning potential. The SHAPE results also demonstrate that the impact of SCD extends to those who care for people living with SCD and significantly affects many aspects of caregivers' lives.

John James OBE, Chief Executive, Sickle Cell Society said: *"The SHAPE UK data shows the stark reality for people living with SCD. This research confirms just how significant the inequalities in SCD care are for people living with the condition in the UK. I'm not surprised most patients and caregivers in the SHAPE survey believe SCD is not seen to be as important as other conditions by society – while this is their lived experience, it was also what we uncovered in our report last year. However, we hope to be able to use this latest data to continue to build our case for improved care for people living with this debilitating disease, as well as those who care for them. We will continue to advocate for better disease management to help improve quality of life and maximise the potential of all those affected by SCD in the UK."*

SHAPE survey patient and caregiver results
SHAPE was administered as an online survey in 10 countries to understand the impact of SCD and quality of life (QoL) of patients and caregivers; of 919 patients

PP-UNP-GBR-2689
November 2022

and 207 caregivers interviewed globally, 151 patients and 30 caregivers were from the UK.

Patients

- The UK SHAPE data showed people with SCD who experienced fatigue/tiredness, headaches or bone aches, either in school or at work, missed an average of **7.9 days of school or work per month** (equivalent to over 3 months missed per year)[1]
- **More than half (56%)** of patients worry they are unable to succeed in education/work because of their SCD[1] and **72% were concerned they would be seen as less competent** in comparison to their colleagues or pupils[1]
- 74% of patients experience **low-mood and/or depression**[1]
- **Fatigue/tiredness** is the most common physical symptom of SCD experienced by 90% of SHAPE survey patient participants in the past year[1]
- Most patients (87%) believe that SCD is **not seen to be as important as other conditions** by society[1]
- 64% of patients reported they had experienced **poor care** in the past from emergency medicine professionals due to a **lack of knowledge about SCD**[1]

Caregivers

- The impact of SCD is not limited to patients, with caregivers stating they lost an average of **6.1 days of school or work per month** (equivalent to almost 2.5 months missed per year)[1] due to caring for someone living with SCD.
- More than half (57%) of caregivers felt that **caring for someone with SCD affected their earning potential**[1] **as well as the ability to attend and succeed at school or work** (53%).[1]
- 80% of UK caregivers have made **sacrifices in the type of jobs** they do because of caring for someone living with SCD[1]
- 80% of UK caregivers have **extra expenses** due to caring for someone with SCD[1]
- Most caregivers (83%) believe that SCD is **not seen to be as important** as other conditions by society[1]

Dr. Baba Inusa, professor and consultant of paediatric haematology, Guy's and St Thomas' NHS Foundation Trust, London and chair of the National Haemoglobinopathy Panel in England said: *"People living with SCD experience progressive and life-threatening complications, including damage to major organs such as the liver, kidneys, lungs and heart which negatively impact quality of life and reduce life expectancy. Results from the SHAPE survey emphasise the importance of understanding patients' needs, and by working together we can help to improve patients' quality of life. It is also important that caregivers are given the support they need, particularly as caring for someone with SCD impacts their own ability to work or attend school and also impacts their earning potential."*

SCD affects approximately 15,000 people in the UK.[2] People living with SCD experience progressive, serious complications and morbidities, including organ

damage, which lead to decreased quality of life and early mortality.[3] Furthermore, economic disadvantages and health inequalities experienced by many patients with SCD can have negative societal impacts in areas such as access to healthcare, education and employment.[4-10]

Mr Nigel Nichols, General Manager of GBT UK said: *"We know that people living with SCD in the UK face ongoing health inequalities; results from the No One's Listening Report and our SHAPE survey confirm this. These inequalities arise due to several factors, including a lack of understanding about the condition, combined with the fact it is more common in people of African or African-Caribbean origin, who tend to have poorer health outcomes and experience higher levels of social deprivation than other ethnicities in the UK. It is therefore crucial that we continue our work to raise awareness of these inequalities, so that we can better understand the impact of SCD across all aspects of peoples' lives, and most importantly so we can work together to make a positive change for those living with the condition and those who care for them."*

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Notes to Editor
About Sickle Cell Disease
It is estimated that more than 100,000 people in the United States,[11] approximately 52,000 people in Europe,[12] up to 100,000 people in Brazil,[13] and millions of people throughout the world have sickle cell disease (SCD).[11] SCD occurs particularly among those whose ancestors are from sub-Saharan Africa; Spanish-speaking regions in the Western Hemisphere (South America, the Caribbean, and Central America); Saudi Arabia; India; and Mediterranean countries such as Turkey, Greece, and Italy.[11] SCD is a lifelong inherited blood disorder that impacts haemoglobin, a protein carried by red blood cells that delivers oxygen to tissues and organs throughout the body.[14] Due to a genetic mutation, individuals with SCD form abnormal haemoglobin known as sickle haemoglobin. When sickle haemoglobin becomes deoxygenated, it polymerises to form rods, which deforms the red blood cells into sickled – crescent-shaped, rigid – cells.[3,15,16] The recurrent sickling process causes destruction of the red blood cells, haemolysis and anaemia (low haemoglobin due to red blood cell destruction), which drives vascular inflammation contributing to blockages in capillaries and small blood vessels (vaso-occlusion) that impede the flow of blood and oxygen delivery throughout the body. Episodes of painful vascular occlusions are commonly referred to as vaso-occlusive crises (VOCs). The diminished oxygen delivery to tissues and organs can lead to life-threatening

complications, including stroke and irreversible organ damage.[3, 15-18] Complications of SCD begin in early childhood and can include neurocognitive impairment, acute chest syndrome, and silent and overt stroke, among other serious issues.[19] Early intervention and treatment of SCD have shown potential to modify the course of this disease, reduce symptoms and events, prevent long-term organ damage, and extend life expectancy.[3]

About Pfizer: Breakthroughs That Change Patients' Lives

At Pfizer, we apply science and our global resources to bring therapies to people that extend and significantly improve their lives. We strive to set the standard for quality, safety and value in the discovery, development and manufacture of health care products, including innovative medicines and vaccines. Every day, Pfizer colleagues work across developed and emerging markets to advance wellness, prevention, treatments and cures that challenge the most feared diseases of our time. Consistent with our responsibility as one of the world's premier innovative biopharmaceutical companies, we collaborate with health care providers, governments and local communities to support and expand access to reliable, affordable health care around the world. For more than 170 years, we have worked to make a difference for all who rely on us. In the UK, Pfizer has its business headquarters in Surrey and is a major supplier of medicines to the NHS. To learn more about our commitments, please visit us at www.pfizer.co.uk or follow us on Twitter (@Pfizer_UK), Facebook (@PfizerUK) and Instagram (@pfizeruk).

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1. GBT Sickle Cell Health Awareness, Perspectives and Experiences (SHAPE) Survey: Final Report. Data on File. (2022) (PP-UNP-GBR-2659)
2. Sickle Cell Society. About Sickle Cell. https://www.sicklecellsociety.org/about-sickle-cell/. Accessed November 2022.
3. Kato GJ, et al. Sickle cell disease. *Nat Rev Dis Primers*. 2018;4:18010.
4. McClish DK, et al. Health related quality of life in sickle cell patients: the PiSCES project. Health Qual Life Outcomes. 2005;3:50.
5. Daniel LC, et al. Lessons Learned From a Randomized Controlled Trial of a Family-Based Intervention to Promote School Functioning for School-Age Children With Sickle Cell Disease. J Pediatr Psychol. 2015;40:1085-1094.
6. Dampier C, et al. Health-related quality of life in adults with sickle cell disease (SCD): a report from the comprehensive sickle cell centers clinical trial consortium. Am J Hematol. 2011;86:203-205.
7. Dampier C, et al. Health-related quality of life in children with sickle cell disease: a report from the Comprehensive Sickle Cell Centers Clinical Trial Consortium. Pediatr Blood Cancer. 2010;55:485-494.
8. Anie KA, et al. Sickle cell disease: Pain, coping and quality of life in a study of adults in the UK. Br J Health Psychol. 2002;7:331-344.
9. Kambasu DM, et al. Health-related quality of life of adolescents with sickle cell disease in sub-Saharan Africa: a cross-sectional study. BMC Hematol. 2019;19:9.
10. Lubeck D, et al. Estimated Life Expectancy and Income of Patients With Sickle Cell Disease Compared With Those Without Sickle Cell Disease. JAMA Netw Open. 2019;2:e1915374.
11. Centers for Disease Control and Prevention. Sickle Cell Disease Data and Statistics (SCD). https://www.cdc.gov/ncbddd/sicklecell/data.html. Accessed November, 2022.
12. European Medicines Agency. https://www.ema.europa.eu/en/medicines/human/orphan-designations/eu3182125. Accessed November, 2020.
13. Ministério da Saúde (Brasil), Protocolo Clínico e Diretrizes Terapêuticas da Doença Falciforme, Feb. 19, 2018

14. National Heart, Lung, and Blood Institute. Sickle Cell Disease. https://www.nhlbi.nih.gov/health-topics/sickle-cell-disease. Accessed November, 2022
15. Rees DC, et al. Sickle-cell disease. *Lancet*. 2010;376(9757):2018-2031. https://pubmed.ncbi.nlm.nih.gov/21131035/ Accessed November 2022
16. Kato GJ, et al. Intravascular hemolysis and the pathophysiology of sickle cell disease. J Clin Invest. 2017;127(3):750-760. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5330745/ Accessed November 2022
17. Caboot JB, et al. Hypoxemia in sickle cell disease: significance and management. *Paediatr Respir Rev*. 2014;15(1):17-23. https://pubmed.ncbi.nlm.nih.gov/24461342/ Accessed November 2022
18. Nader E, et al. The Red Blood Cell-Inflammation Vicious Circle in Sickle Cell Disease. *Front Immunol*. 2020 Mar 13;11:454. https://pubmed.ncbi.nlm.nih.gov/32231672/ Accessed November 2022
19. Kanter J, et al. Management of sickle cell disease from childhood through adulthood. *Blood Rev*. 2013 Nov;27(6):279-87. https://pubmed.ncbi.nlm.nih.gov/24094945/ Accessed November 2022