

Understanding the health-care experiences of people with sickle cell disorder transitioning from paediatric to adult services: This Sickle Cell Life, a longitudinal qualitative study

Alicia Renedo,¹ Sam Miles,¹ Subarna Chakravorty,²
Andrea Leigh,³ John O Warner^{4,5} and
Cicely Marston^{1*}

¹Department of Public Health, Environments and Society, Faculty of Public Health and Policy, London School of Hygiene & Tropical Medicine, London, UK

²Department of Paediatric Haematology, King's College Hospital, London, UK

³University College London NHS Hospitals Foundation Trust, London, UK

⁴National Heart and Lung Institute, Imperial College London, London, UK

⁵Collaboration for Leadership in Applied Health Research and Care for Northwest London, Imperial College London, London, UK

*Corresponding author Cicely.Marston@lshtm.ac.uk

Declared competing interests of authors: Subarna Chakravorty reports receiving honoraria for taking part in advisory boards for Novartis Pharmaceuticals UK Ltd (London, UK) and an honorarium from Novartis Pharmaceuticals UK Ltd for participating in the development of a national consensus guideline document for the management of health-care transition services in sickle cell disorder. John O Warner reports acting as a principal investigator and a scientific advisory board member (2005 to present) for trials of infant milk formulae for the prevention of allergy (Danone SA, Paris, France/Nutricia, Zoetermeer, the Netherlands); acting as an investigator and a scientific advisory board member (2008 to present) for trials of an environmental control system for allergic conditions (Airsonett AB, Ängelholm, Sweden); bursaries for lectures at conferences for Danone SA/Nutricia, Airsonett AB, UCB Pharma (Brussels, Belgium), Novartis Pharmaceuticals UK Ltd and Allergy Therapeutics plc (Worthing, UK); acting as medical advisor (1993 to present) for the Anaphylaxis Campaign (www.anaphylaxis.org.uk); membership of the scientific advisory board (2017 to present) for a UK government Home Office enquiry into potential toxic health hazards from the Grenfell Tower fire; and acting as academic theme lead (2014–19) for the Collaboration for Leadership in Applied Health Research and Care for Northwest London (until 30 September 2019).

Published November 2020

DOI: [10.3310/hsdr08440](https://doi.org/10.3310/hsdr08440)

Scientific summary

This Sickle Cell Life, a longitudinal qualitative study

Health Services and Delivery Research 2020; Vol. 8: No. 44

DOI: [10.3310/hsdr08440](https://doi.org/10.3310/hsdr08440)

NIHR Journals Library www.journalslibrary.nihr.ac.uk

Scientific summary

Background

Transition from paediatric to adult health services causes problems worldwide, particularly among young people living with long-term conditions. These problems affect individuals' health and future life, and improvements in transition are urgently needed. Health-care transition spans the period from early adolescence to adulthood and continues until the individual adjusts to adult health-care services. Transitioning is a time of increased vulnerability, when young people with long-term conditions face multiple demands from diverse social contexts beyond health care, including the need for increased competency in self-management of their condition and societal expectations about the need to develop adult behaviours.

Sickle cell disorder is a long-term condition for which transition to adult health-care services is particularly challenging. Sickle cell disorder is an inherited blood disorder characterised by abnormal haemoglobin that results in sickle-shaped red blood cells. It leads to anaemia, jaundice, fatigue, acute and chronic pain, and extreme tiredness. Episodic pain episodes are a constant threat for people with sickle cell disorder. They require timely medical treatment and can become more frequent with increasing age. Pain episodes can be excruciating and sometimes evolve into acute chest syndrome, stroke and/or severe infection.

Listening to young people's voices – and acting on what they say – is crucial to develop better and more youth-friendly health-care services. Young people's voices are not yet sufficiently addressed in health-care transition research and there is little research examining how experiences of clinical care beyond specialist services or how young people's wider social context affect the transition process. Understanding young people's experiences across the health-care system during transition, as well as how aspects of their social life affect transition, is crucial to inform the support that young people need to prepare them for adult health services and adult life.

Objectives

To inform better service provision by improving understanding of experiences of young people with sickle cell disorder as they transition between child and adult health-care services and how this health-care transition interacts with their wider social contexts.

Methods

We used a longitudinal qualitative study design. Data collection included 80 interviews in 2016–17 with young people (aged 13–21 years) with sickle cell disorder recruited across two cities in England. We interviewed 48 participants (30 females and 18 males). We conducted 27 one-off interviews and 53 repeated interviews (i.e. interviews conducted two or three times over 18 months). We additionally interviewed 10 sickle cell disease specialist health-care providers.

We conducted repeated interviews with younger participants to capture their experiences during the process of transitioning into adult services and changes over time. This helped to capture the evolving experience of living with sickle cell disorder. We conducted single interviews with older participants who had already been transferred to adult care. Transition is a dynamic process from when transition is introduced in health-care services through to the post-transfer period when young people are

adjusting to adult services. We included young participants (aged 13–15 years) who were experiencing the introduction to transition in sickle cell disease clinics and also older participants who had already been transferred to adult services to capture their retrospective accounts and their current experiences of adult services. We conducted 10 interviews with sickle cell disease specialist health-care providers about their views and experiences of providing care for patients with sickle cell disorder undergoing health-care transition.

We used an inductive analytical approach, combining elements of grounded theory and thematic analysis.

This is a co-produced study. We worked closely with non-clinical, non-academic sickle cell disorder experts (patient advocates) throughout the study from its inception to research design, analysis and dissemination.

Results

A key challenge for young people with sickle cell disorder during transition is the difficulty of being heard. Their knowledge of their body and of how the condition affects them is frequently disregarded in interactions with others. Their expertise is ignored during unplanned visits in accident and emergency departments and on general hospital wards when they are admitted as inpatients for acute exacerbations, mostly with painful episodes.

Young people struggle with health-care transitions for several key reasons. First, there are tensions between the expectations of them as adult patients (e.g. being independent, engaging in 'good' self-care) and the realities of the disabling environments they have to navigate. On the one hand, health-care providers, parents and carers demand that the young person learn more about their condition and body, and act responsibly to stay healthy. On the other hand, when young people attempt to practise these new behaviours and use their patient expertise, for instance by making requests about the care they receive, they are often disregarded, questioned and made to feel invisible.

Young people identified the following obstacles to the receipt of high-quality unplanned hospital care:

- Their pain relief needs are ignored (e.g. they experience delays in receiving medication for pain relief).
- They are ignored when they ask for help with basic bodily care (e.g. toileting).
- Non-specialist health-care staff do not know enough about sickle cell disorder.
- They experience a lack of voice and barriers to involvement in decisions about their care because non-specialist health-care staff do not recognise the patient's expertise in their own condition and needs.
- Adult wards are unwelcoming and young people want to avoid them.
- Non-specialist staff appear sceptical about young people's voices and needs. This scepticism is communicated directly (by questioning young people) or expressed more obliquely by staff delaying pain treatment. Several participants talked about occasions when they felt that staff suspected them of drug-seeking.

Negative health-care experiences during the period of transition create uncertainty about quality of care in non-specialist hospital settings and can adversely affect young people's health-care seeking. For young people with sickle cell disorder, the period of transition involves increasing awareness of the shortcomings of unplanned non-specialist hospital care and the obstacles to being considered legitimate patient experts in these health-care contexts. Young people learn that their condition can be misunderstood by non-specialist staff, who do not always listen to their requests, respond to their needs or involve them in decisions about their care. They develop mistrust in non-specialist hospital

care and try to disengage as much as possible from these services by trying to find ways to stay out of hospital as much as possible. As a result of these experiences, transition involves young people pushing themselves into self-reliance and into learning how to call on their expertise in their own body and their own health condition to stay out of hospital. Even though our participants were aware that avoiding visiting hospital could be dangerous for their health, going to hospital during pain episodes was described as a last resort.

During transition, young people with sickle cell disorder also encounter barriers to enacting their patient expertise when interacting with others in their social context (e.g. friends, peers and adults at school/work). Participants talked about others questioning or undermining their reports of acute pain or sickle cell disorder-related fatigue. They also told us of the opposite happening: others could be overprotective or over-react to their signs of pain and ignore participants' explanations that the situation was not an emergency.

In their social lives outside hospital, participants resorted to social silencing, avoiding disclosing their condition to others because they expected that others would not listen properly or would not understand. Social silencing also meant that they did not ask for support when they needed it and avoided mentioning sickle cell disorder to explain why they were fatigued or why they arrived at school late. Social silencing helped protect participants from having to constantly explain sickle cell disorder to others who they expected would not understand. It also helped younger participants who struggled to explain sickle cell disorder to others.

Young people's social lives interact with their health transitions and their movement into adult patienthood. Young people with sickle cell disorder encounter difficulties when trying to develop adult identities that help them to achieve life goals while simultaneously staying healthy. They can struggle to develop a coherent sense of adult self because they are caught between the conflicting demands of being responsible patients, acting to preserve health, but also self-actualising adults. Identity development during transitions involve relentless self-disciplining and self-monitoring in different aspects of their lives. Young people see themselves as being in constant need of improvement to reconcile the plural demands on them (e.g. to produce bodily efficacy and stay healthy while simultaneously working hard to become competent in other areas of their life, such as education and work, to become the type of productive adult that others wish them to be and that they themselves wish to be). Fulfilling educational demands is difficult for young people with sickle cell disorder who have to spend time in hospital or who are in pain or have to rest regularly to avoid pain episodes. At the same time, health-care staff and parents push young people to take individual responsibility and become competent at sickle cell disorder self-management. The difficulty of trying to develop self-actualising adult identities while also being a disciplined adult patient plays out in stigmatising identities (e.g. self or others' characterisations of the young person with sickle cell disorder as lazy).

Relational dimensions of the sickle cell disorder experience are crucial – that is, how the condition is lived and accounted for through and against significant others, particularly family members. Participants seemed to prioritise the ways their sickle cell disorder affects others over how it affects themselves. For instance, they talked about wanting to get better to avoid inconveniencing others and how they try to hide their pain to avoid worrying their parents. Young people's perceptions of the impact of sickle cell disorder on significant others and the feeling of being a burden to them add stress to an already painful and unpredictable condition. Moving into adulthood involves learning how to keep sickle cell disorder private, managing symptoms alone and disguising pain. Internalising illness experiences becomes part of young people's repertoire of self-management tactics as they move into adulthood.

Conclusions

Our study emphasises the need for services to become more person centred and respectful of patient expertise. Health-care providers should solicit and act on young people's voices to ensure that they are involved in shaping their own health care. If young people are prevented from using transition skills (e.g. self-management and self-advocacy) or are treated by staff with inadequate knowledge of their condition then they may well lose trust in services and disengage from them, potentially compromising their health.

The care young people receive across the health-care system, including during interactions with staff, must help young people take responsibility for their own health and advocate for their own needs. Our findings show that health-care services outside specialised care are failing young people with sickle cell disorder. It is vital to work with staff outside specialist services to train them in transitional care, sickle cell disorder care and adolescent health, and to help them develop compassionate care and communication skills. Engaging with young people and listening to their voices is crucial to ensure that they can influence their own health care. Staff should be supported to recognise young people's status as experts in their own body and condition and must recognise young people's voices as legitimate. It is also important to address negative stereotyping to ensure that people with sickle cell disorder are able to access opioids for pain relief when they need them.

Families and carers may be able to support young people with internal conflicts during transitions, for instance in relation to developing adult identities or attempts to protect significant others. Work with families and carers could include helping to facilitate open conversations at home about the additional stress young people experience when they try to protect others from the suffering and impact of their condition.

Health-care transition support work could also work with schools or other institutions outside services to create supportive spaces that might help young people to use their patient expertise to protect their health while still consolidating self-actualising adult identities that could help young people achieve their life goals. Substantial work is needed in this wider social context to raise awareness about the condition, educate the public and address the stigma associated with sickle cell disorder-related fatigue.

Funding

This project was funded by the National Institute for Health Research (NIHR) Health Services and Delivery Research programme and will be published in full in *Health Services and Delivery Research*; Vol. 8, No. 44. See the NIHR Journals Library website for further project information.

Health Services and Delivery Research

ISSN 2050-4349 (Print)

ISSN 2050-4357 (Online)

This journal is a member of and subscribes to the principles of the Committee on Publication Ethics (COPE) (www.publicationethics.org/).

Editorial contact: journals.library@nihr.ac.uk

The full HS&DR archive is freely available to view online at www.journalslibrary.nihr.ac.uk/hsdr. Print-on-demand copies can be purchased from the report pages of the NIHR Journals Library website: www.journalslibrary.nihr.ac.uk

Criteria for inclusion in the *Health Services and Delivery Research* journal

Reports are published in *Health Services and Delivery Research* (HS&DR) if (1) they have resulted from work for the HS&DR programme, and (2) they are of a sufficiently high scientific quality as assessed by the reviewers and editors.

HS&DR programme

The HS&DR programme funds research to produce evidence to impact on the quality, accessibility and organisation of health and social care services. This includes evaluations of how the NHS and social care might improve delivery of services.

For more information about the HS&DR programme please visit the website at <https://www.nihr.ac.uk/explore-nihr/funding-programmes/health-services-and-delivery-research.htm>

This report

The research reported in this issue of the journal was funded by the HS&DR programme or one of its preceding programmes as project number 13/54/25. The contractual start date was in April 2015. The final report began editorial review in December 2019 and was accepted for publication in June 2020. The authors have been wholly responsible for all data collection, analysis and interpretation, and for writing up their work. The HS&DR editors and production house have tried to ensure the accuracy of the authors' report and would like to thank the reviewers for their constructive comments on the final report document. However, they do not accept liability for damages or losses arising from material published in this report.

This report presents independent research funded by the National Institute for Health Research (NIHR). The views and opinions expressed by authors in this publication are those of the authors and do not necessarily reflect those of the NHS, the NIHR, NETSCC, the HS&DR programme or the Department of Health and Social Care. If there are verbatim quotations included in this publication the views and opinions expressed by the interviewees are those of the interviewees and do not necessarily reflect those of the authors, those of the NHS, the NIHR, NETSCC, the HS&DR programme or the Department of Health and Social Care.

© Queen's Printer and Controller of HMSO 2020. This work was produced by Renedo *et al.* under the terms of a commissioning contract issued by the Secretary of State for Health and Social Care. This issue may be freely reproduced for the purposes of private research and study and extracts (or indeed, the full report) may be included in professional journals provided that suitable acknowledgement is made and the reproduction is not associated with any form of advertising. Applications for commercial reproduction should be addressed to: NIHR Journals Library, National Institute for Health Research, Evaluation, Trials and Studies Coordinating Centre, Alpha House, University of Southampton Science Park, Southampton SO16 7NS, UK.

Published by the NIHR Journals Library (www.journalslibrary.nihr.ac.uk), produced by Prepress Projects Ltd, Perth, Scotland (www.prepress-projects.co.uk).

Editor-in-Chief of *Health Services and Delivery Research* and NIHR Journals Library

Professor Ken Stein Professor of Public Health, University of Exeter Medical School, UK

NIHR Journals Library Editors

Professor John Powell Chair of HTA and EME Editorial Board and Editor-in-Chief of HTA and EME journals. Consultant Clinical Adviser, National Institute for Health and Care Excellence (NICE), UK, and Professor of Digital Health Care, Nuffield Department of Primary Care Health Sciences, University of Oxford, UK

Professor Andrée Le May Chair of NIHR Journals Library Editorial Group (HS&DR, PGfAR, PHR journals) and Editor-in-Chief of HS&DR, PGfAR, PHR journals

Professor Matthias Beck Professor of Management, Cork University Business School, Department of Management and Marketing, University College Cork, Ireland

Dr Tessa Crilly Director, Crystal Blue Consulting Ltd, UK

Dr Eugenia Cronin Senior Scientific Advisor, Wessex Institute, UK

Dr Peter Davidson Consultant Advisor, Wessex Institute, University of Southampton, UK

Ms Tara Lamont Senior Scientific Adviser (Evidence Use), Wessex Institute, University of Southampton, UK

Dr Catriona McDaid Senior Research Fellow, York Trials Unit, Department of Health Sciences, University of York, UK

Professor William McGuire Professor of Child Health, Hull York Medical School, University of York, UK

Professor Geoffrey Meads Emeritus Professor of Wellbeing Research, University of Winchester, UK

Professor John Norrie Chair in Medical Statistics, University of Edinburgh, UK

Professor James Raftery Professor of Health Technology Assessment, Wessex Institute, Faculty of Medicine, University of Southampton, UK

Dr Rob Riemsma Reviews Manager, Kleijnen Systematic Reviews Ltd, UK

Professor Helen Roberts Professor of Child Health Research, UCL Great Ormond Street Institute of Child Health, UK

Professor Jonathan Ross Professor of Sexual Health and HIV, University Hospital Birmingham, UK

Professor Helen Snooks Professor of Health Services Research, Institute of Life Science, College of Medicine, Swansea University, UK

Professor Ken Stein Professor of Public Health, University of Exeter Medical School, UK

Professor Jim Thornton Professor of Obstetrics and Gynaecology, Faculty of Medicine and Health Sciences, University of Nottingham, UK

Professor Martin Underwood Warwick Clinical Trials Unit, Warwick Medical School, University of Warwick, UK

Please visit the website for a list of editors: www.journalslibrary.nihr.ac.uk/about/editors

Editorial contact: journals.library@nihr.ac.uk