Paediatric to adult transition care for patients with sickle cell disease: a global perspective

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Sickle cell disease is a life-threatening inherited condition designated as a public health priority by WHO. Increased longevity of patients with sickle cell disease in high-income, middle-income, and low-income countries present unprecedented challenges for all settings; however, a globally standardised solution for patient transition from paediatric to adult sickle cell disease health care is unlikely to address the challenges. We established a task force of experts from a multicountry (the USA, Europe, Middle East, and Africa) consortium. We combined themes from the literature with viewpoints from members of the task force and invited experts to provide a global overview of transition care practice, highlighting barriers to effective transition care and provide baseline recommendations that can be adapted to local needs. We highlighted priorities to consider for any young person with sickle cell disease transitioning from paediatric to adult health care: skills transfer, increasing self-efficacy, coordination, knowledge transfer, linking to adult services, and evaluating readiness (the SICKLE recommendations). These recommendations aim to ensure appropriate benchmarking of transition programming, but multisite prospective studies are needed to address this growing public health need.

Lancet Haematol 2020; 7: e329–41

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Introduction

Sickle cell disease is one of the most common life-threatening inherited conditions worldwide and has been designated a public health priority by WHO.[1] The annual number of babies born with either homozygous or double heterozygous forms of sickle cell disease is estimated to be more 400 000 and the majority (>80%) of these babies are born in low-income and middle-income settings, especially sub-Saharan Africa.[2] In sub-Saharan Africa where there are substantially fewer resources, childhood mortality prevalence is substantially higher compared with high-income counties (HICs). In the late 1970s, the probability of children with sickle cell disease dying by the age of 5 years was reported to be more than 90% in some rural communities of northern Nigeria.[3] However as medical advances filter down to communities across the globe, the prognosis for sickle cell disease in sub-Saharan Africa has improved because of increasing access to health care and lower exposure to infectious diseases, and now 50% of children with sickle cell disease will survive beyond 10 years of age.[4] 50 years ago in the USA, less than 50% of children with sickle cell disease were reported to survived until adulthood.[5] However, data show that patient survival has improved and more than 95% of babies born with sickle cell disease in the USA and the UK will survive into adulthood.[6,7] These improvements are mainly because of better screening and preventative treatment processes, including prophylactic penicillin, vaccinations, and therapies, such as hydroxyurea, in addition to improved supportive care.[1] The transformation of sickle cell disease from a disease of childhood into a chronic, lifelong condition is unprecedented,[8,9] and it imposes increasing new challenges as those with the disease grow into adulthood.

Because children with sickle cell disease are now expected to live longer, management of their transition from childhood to adulthood and the shift from paediatric to adult services has become an essential step for health-care professionals. Care transition is defined by Blum and colleagues[10] as "the purposeful, planned movement of adolescents and young adults from child-centred to adult-oriented health care systems." This transition process prepares young people for the actual transfer to adult care.[11] The transition period can be a difficult time not only for the adolescent, but also their families and the health-care providers working within the health-care system. A family's ability to assist their adolescent to cope with and effectively manage their disease substantially influences the young person's behaviours, adherence to treatment, and outcomes.[12] The availability of knowledgeable adult-oriented health-care providers has not kept pace with the rapidly growing population of adults with sickle cell disease.[13]

The movement from paediatric to adult health services poses important challenges and is widely acknowledged as a period of high risk, incurring the highest usage of emergency services.[11–13] In England, sickle cell disease related hospital admissions increased by more than 50% between 2001 and 2010 with the highest number of emergency admissions reported in patients who had recently transitioned from paediatric to adult care.[14] This is corroborated by data from the Dallas Newborn Cohort study in the United States,[15] which identified that the period with the highest mortality for patients with sickle cell disease is now around the time of transition and transfer. This increase in morbidity and mortality and acute care utilisation for use during the transition years is because of a combination of factors including the small number of adult oriented practitioners to provide care, an increase in certain comorbidities (eg, renal and cardiac disease, iron overload, and silent cerebral infarcts) as patients age, and behavioural and cognitive challenges that influence adherence.[12,15,16] Young people who have a high reliance on emergency services for

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	Institutions using the method	Advantages	Challenges
Individual curriculum	Got Transition, The National Alliance to Advance Adolescent Health, Washington DC, USA and Sickle Cell Center, Virginia Commonwealth University, Richmond, VA, USA	Tailored education and programming	Possible repetition and gaps
Group curriculum	Children's Healthcare of Atlanta, Richmond, VA, USA	Peer support	Hard to schedule a joint clinic meeting, peer support is not the most reliable, and a session leader must be trained and retained
Online curriculum	St Jude's Research Hospital, Memphis, TN, USA and Children's National, Memphis, TN, and Washington DC, USA	Convenience and accessibility	Limited opportunities to give feedback
Smartphone app-based curriculum	St Jude's Research Hospital, Memphis, TN, USA	Convenience and accessibility	Limited opportunities to give feedback
Peer mentoring	University of Illinois at Chicago, Chicago, IL, USA	Peer support	Mentors have to be trained and retained and the quality of peer support is not reliable
Group care, including family support	University of Illinois at Chicago, Chicago, IL, USA and University of California San Francisco Benioff Children's Hospital Oakland, Oakland, California, USA	Peer and parent-to-parent support	Hard to schedule a joint clinic meeting, peer support is not the most reliable, and a session leader must be trained and retained
Colocated care	Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, USA, University of California San Francisco Benioff Children's Hospital Oakland, Oakland, California, USA, University of Connecticut Health, Farmington, CT, USA, and Center for Sickle Cell Disease, St Jude-University of Tennessee Health Science Center, Memphis, TN, USA	Visible paediatric and adult collaboration	It is difficult to organise collaboration between paediatric and adult care specialists at a single site of care
Structured questionnaires to guide consultations around transition	Ready Steady Go, University of Southampton, UK	No additional staff required, and it is a very flexible age of implementation	The programme is not specific to sickle cell disease
Patient passport	Guy's and St Thomas NHS Trust, London, UK	Individualised education curriculum	Requires dedicated staff for care transition and the process is resource intense

Table 1: **Examples of different care transition programme models currently in use**

their care incur more days in hospital and substantially higher overall health-care costs.[15–19] Successful transition from paediatric to adult health-care services is correlated with better outcomes for patients with sickle cell disease because of better adherence, medication management, and self-care.[20–22] As such there is an urgent need for improvement in sickle cell disease care, particularly relating to the time of transition.[22–25]

See **Online** for appendix

This Review aims to combine themes from the literature with viewpoints from a task force of practitioners to provide an overview of global practice, highlight key barriers to effective transition, and provide a global consensus on priorities for practice.

Task force constitution

This Review was commissioned through the African Research and Innovative Initiative for Sickle Cell Education (ARISE) consortium and invited experts on transition in sickle cell disease. The experts were initially selected based on their publications on the subject of transition and also to represent key regions of the world, including Africa, the USA, the EU, and the Middle East. Later, other leading experts from Brazil, Democratic Republic of Congo, India, and Saudi Arabia were approached for their contributions.

ARISE is an EU funded interagency and multi-disciplinary staff exchange programme between researchers, involving a total of 20 institutions from the EU, Africa, Lebanon, and the USA to foster the sharing of best practices in newborn screening, diagnosis, and treatment of sickle cell disease aimed to improve overall disease outcomes. This consortium uses implementation science strategies to foster partnerships and the establishment of a patient database, registries, and sustainable services for people with sickle cell disease. The legacy of ARISE (appendix) brings together experts in sickle cell disease clinical care, epidemiology, basic science, genetics, and the social sciences to foster excellent clinical and research collaborations. ARISE activities have the potential to become a template for future international collaboration to enhance health services and research in Africa. We are in the process of establishing a consortium to strengthen research capacity through innovative staff exchange using different objectives including the work package for the development of best practice in transition from paediatric to adult care.

Global practices on paediatric to adult transition care

The boundaries between paediatric care and adult care, such as the patient's age at transfer, differ substantially between different countries and regions of the world, thereby making transition a cultural construct with respect to autonomy and responsibility.[9,15–17,20–27] In many African countries the age at transfer ranges from

12 to 15 years.[18,20] The situation in the countries around the Persian Gulf appears to be even more challenging because the age limit for admission to a paediatric ward is 12 years.[21] Therefore, beyond this age, an adolescent must be admitted to an adult hospital and treated by adult physicians and adult trained nurses.[21] In the USA, the age of transfer of care varies between states and centres, but usually occurs between 18 years and 21 years of age,[22,27] with transition first discussed when the patient is, on average, 15 years old.[19] In Canada, transfer of care occurs typically between 17–19 years of age.[23] In the UK it is common practice to start planning for adulthood from the age of 13–14 years, although, again, the national guidelines clearly state the transition planning is developmentally appropriate and takes into account each young person's capabilities, needs, and hopes for the future.[24,28] The point of transfer of care is not based on a rigid age threshold, but in practice usually occurs at 16–18 years, with some exceptional patients remaining in the paediatric-care system beyond the age of 18 years.

Aside from age at transfer of care, where transfer occurs also varies greatly and the structure of the health-care system also significantly effects transition.[26,29,30] Sometimes the specialist care for sickle cell disease has a separate paediatric centre provider from the adult provider, but for others the haematological provision specialty spans all ages.[25] Transfer of care can be more challenging to successfully complete if the paediatric service stands alone because transition should be a shared task between the paediatric and adult health-care provider.[30] Furthermore, the use of multiple health-care providers by young adults complicates this process and makes consistent patient education and transition planning difficult.[27,29,30]

High-income country settings

In HICs, children and adolescents diagnosed with chronic medical conditions are expected to experience their health-care transition in a systematic and structured process, often within transition programmes (table 1).[26] However, even HIC settings are not exempt from the challenges of resource limitations and poor infrastructure to meet the needs of adolescents and young adults; resulting in the development and testing of a plethora of strategies to improve the transition process in different centres and countries. In addition to local transition programmes, many HIC settings have developed national guidance on transition. In 2016, the American Society of Hematology helped to design a toolkit to standardise and streamline the transition process for patients with life-long haematological disease, including sickle cell disease. In 2011, the American Academy of Paediatrics, American Academy of Family Physicians, and American College of Physicians[27] established an algorithm of the key aspects of health-care transition. This algorithm was expanded with clinical tools and called, the Six Core Elements of Health Care Transition, which included the incorporation of a transition policy, transition tracking and monitoring, assessment of transition readiness, the actual transfer to adult-centred care, and transition completion and ongoing adult-centred care. The Got Transition programme[29] developed by the National Alliance to Advance Adolescent Medicine is a national initiative in the USA that aims to improve health-care transition on the basis of the Six Core Elements of Health Care Transition, with resources that could be customised based on the type of practice. The 2011 clinical report was updated in 2018 to include recommendations on health-care system infrastructure, initiatives for training providers about caring for transitioning individuals, and payment mechanisms for transition-related care.[27]

In Canada, there have been trials of transition coordinators who are involved in the care of patients from the paediatric period through transfer and into adult services.[23,30] The UK has produced national guidelines regarding the transition process for children with sickle cell disease that highlight the importance of the multidisciplinary team and community services and the use of a patient passport to assess individual patient readiness based on their knowledge of their disease and self-efficacy.[31] In France there is no standardised transition programme, but French studies have emphasised the significance of care continuity, good communication, multidisciplinary teams, and have highlighted the importance of addressing parent and caregiver anxieties in relation to the transition process.[32]

The model of health care in the USA means that once a patient is deemed an adult insurance policies can be effected; as a result, finances are another major barrier to care that must be considered.[12,33] The effect of health-care resource restrictions in HICs disproportionately affects families of ethnic minorities and those with a high index of deprivation.[26] Worldwide, a few studies have explored transition perspectives and have found similar challenges, including concern about family burden and social stigma.[18,34,35] The overall approach to sickle cell disease management, especially in Africa and India where the disease burden is highest, must develop the strategies to deal with the life-course issues from Newborn Screening through childhood and adolescents (table 1).

Low and middle income country settings

Sub-Saharan Africa bears more than 80% of the global sickle cell disease burden, but many of the countries in which the disease is most prevalent do not have the resources needed for the complex care required by these patients.[34,36–38] Although the largest burden of disease is reported to be in Nigeria, India, and Democratic Republic of the Congo, there are no formalised transition programmes or guidelines for adolescents and young people who need to move into adult care.[36] Many countries are unprepared for the rapidly increasing numbers of

Panel 1: **Global standards for quality health-care services for adolescents**

Standard 1: Adolescents' health literacy
The health facility implements systems to ensure that adolescents are knowledgeable about their own health, and they know where and when to obtain health services.

Standard 2: Community support
The health facility implements systems to ensure that parents, guardians, and other community members and community organisations recognise the value of providing health-care services to adolescents, and support the provisions and use of services by adolescents.

Standard 3: Appropriate package of services
The health facility provides a package of information, counselling, diagnostic, treatment, and care services that fulfils the needs of all adolescents. Services are provided in the facility and through referral linkages and outreach.[1]

Standard 4: Providers' competencies
Health-care providers have the technical competence required to provide effective care services to adolescents. Both health-care providers and support staff respect, protect, and fulfil adolescents' rights to information, privacy, confidentiality, non-discrimination, non-judgemental attitude, and respect.

Standard 5: Facility characteristics
The health facility has convenient operating hours, a welcoming and clean environment, and maintains privacy and confidentiality. It has the equipment, medicines, supplies, and technology needed to ensure effective service provision to adolescents.

Standard 6: Equity and non-discrimination
The health-care facility provides quality services to all adolescents irrespective of their ability to pay, age, sex, marital status, education level, ethnic origin, sexual orientation, or other characteristics.

Standard 7: Data and quality improvement
The health facility collects, analyses and uses data on service use and quality of care, disaggregated by age and sex, to support quality improvement. Health facility staff are supported to participate in continuous quality improvement.

Standard 8: Adolescents' participation
Adolescents are involved in the planning, monitoring, and evaluation of health services and in decisions regarding their care and certain appropriate aspects of service provision.

adults now living with sickle cell disease and these patients are often still cared for by paediatric services well into adulthood.[38] Current practice in Ghana is for the transfer of care to occur by the time the patient is 14–15 years old, with transitioning discussed from 12 years of age.[18] In South Africa, although the transitioning usually begins after the age of 12 years, there is local variation from centre to centre.[21] Personal communications with specialists from Democratic Republic of the Congo (Leon Tshilolo, Monkole Institute, Kinshasa, Democratic Republic of the Congo), India (Priyanka Samal, Department Clinical Haematology and Stem Cell Transplant Institute of Medical Sciences and SUM Hospital Bhubaneswar, India), and the Middle East (Hatoon Ezzat Adult Hemoglobinopathy programme of British Columbia, Canada and Head of Sickle programme, Ministry of Health, Saudi Arabia) confirmed that there is no formal transition programming in place. However, in the Arab states of the Persian Gulf, such as Bahrain, the care transition policy is that children aged 14 years and older are admitted to adult wards, but they remain under the care of the paediatric consultant. In Saudi Arabia a task force has been set up to review sickle cell service provision, including transition. In Brazil there is no formal transition plan, but in some institutions the service providers remain the same team across the patient's life span (Jane Hankins, St Jude's Children's Research Hospital Memphis, TN, USA). Nevertheless, ongoing discussions are taking place in these settings and are at different stages to adapt a workable plan for transition.

In lower and middle income countries (LMICs) the efforts to make up for large health-care personnel gaps has benefitted from the global strategy known as task sharing, task shifting (TSTS). Tasks usually done by highly qualified professionals, such as physicians, are passed on to allied health professionals (eg, community health workers) after adequate training but under supervision. This method of improving human resources for health care has been used for decades for improving coverage and quality of care in HIV, tuberculosis, mental health, cardiovascular disease, and diabetes care in the communities of many LMICs since the WHO Alma-Ata declaration, which identified primary health care as key to attaining the goal of Health For All. Countries in Asia and Africa along with Brazil and other LMICs have different designations for health personnel involved in the TSTS health model, including non-physician clinicians, nurse assistants, pharmacy technicians, community health extension workers (CHEWs), community health extension officers, and traditional birth attendants.

CHEWs are mid-level health workers admitted into schools of health technology with the same educational entry qualifications as nurses and trained for 1–3 years to treat minor ailments, give first aid, develop health awareness, facilitate health education, work within multidisciplinary teams, and refer patients for more specialised care in referral centres. In sub-Saharan Africa, these specially trained CHEWs are well suited to reduce the effect of unequal human resources on health and have supported the 2014 WHO guided National TSTS policy. CHEWs are supported by well designed manuals, protocols, checklists, and supervision, which makes it easy to audit and retrain staff, as designed in the TSTS policy. Because they are recruited from local populations and often speak the local languages CHEWs are a locally acceptable workforce, with access to almost every household in suburban and rural areas, and they can be deployed to ensure complete coverage. Therefore, CHEWs are in a position to empower individuals with chronic diseases and their families. It is more cost-effective and efficient to train CHEWs and clinical medical officers to aid transition planning with patients and their families. CHEWs could be a cost effective and efficient resource for education and strengthening

	Process	Output
The governance; structure of the facility; includes adolescents	The health-care facility carries out regular activities to identify adolescents' expectations about the service,[2] to assess their experience of care, and involves adolescents in the planning, monitoring, and evaluation of health-care services	Adolescents are involved in planning, monitoring, and evaluation of health services
There is a policy in place to engage adolescents in service planning, monitoring, and evaluation	Health-care providers provide accurate and clear information on the medical condition, and disease management, and treatment options, and explicitly take into account the adolescent's decision on the preferred option and follow-up actions	Adolescents are involved in decisions regarding their own care
Health-care providers are aware of laws and regulations that govern informed consent, and the consent process is clearly defined by facility policies and procedures in line with laws and regulations	The health facility carries out activities to build adolescents' capacity in certain aspects of health-service provision	Adolescents are involved in certain aspects of health service provision

Table 2: **Measurable criteria to ensure the involvement of adolescents in the planning, monitoring, and evaluation of health-care services and in decisions regarding their care**

organisational linkages required for optimal transition for paediatric to adult sickle cell disease care.

A 2015 WHO position paper[38] stated that both HIC and LMICs need to consider adopting a standard-driven approach to improve the quality of adolescent health. Following a multistage process that involved systematic reviews of published and non-published data on the health of adolescents, a working group of experts and technical staff was assembled to assess the available data and subject it to further peer review to refine the recommendations. Panel 1 shows the final eight recommended standards that were then field tested in 25 low-income, middle-income, and high-income countries in Asia, Africa, South America, and Europe. Table 2 describes measurable criteria for the WHO standard 8 on adolescents' participation in their care provision.[39]

"The primary intention of the standards is to improve the quality of care for adolescents in government health-care services; however, they are equally applicable to facilities run by non-government organisations and those in the private sector. The ultimate purpose of implementing the standards is to increase adolescents' use of services and, thus, to contribute to better health outcomes".[38]

Developing benchmarking strategies for health-care transition

A review, published in 2015, concluded that "currently, there is no established metric for successful health care transition".[38] However, the urgent need for improved transition practices from paediatric to adult services in sickle cell disease compels the implementation of transition interventions now. While awaiting consensus on metrics for success, health-care transition can be improved by careful selection of best practices and multifaceted evaluation of what works in specific settings. Here, we Review common themes from the literature regarding what contributes to successful care transition and highlight six strategies consideration when developing a transition plan for young people with sickle cell disease (the SICKLE recommendations).[38–47]

Skills training to empower young people

Patient empowerment requires an individual to take care of their own condition and make choices about their care from the options identified by the health-care provider. Patient empowerment is fundamental to prepare children for the change from a paediatric model of care where parents make most decisions to an adult model of care where the patient ultimately takes full responsibility for decision-making. Many patients with sickle cell disease and clinical experts acknowledge and highlight the need for adolescents and young adults to be prepared to take responsibility for their disease management before the transfer of care process begins.[42] Health-care providers working with sickle cell disease are in a prime position to make use of health-promoting strategies and empower young people to advocate for themselves. This is an opportunity to offer developmentally appropriate training on how to self-manage symptoms, such as pain, and how to access professional advice. It is important to address the role of the family and parents continuing this empowerment at home. Young adults who have developed skills during childhood to facilitate their health-care needs are much more likely to maintain positive health habits.[43]

Ensuring that adolescents with sickle cell disease have the skills to navigate adult health-care systems is an equally important aspect for successful transfer to adult care.[44] In addition to health-related skills, young people transitioning from paediatric to adult care need to develop their independent living skills, such as money management and vocational skills.[44] The process of teaching young people to self-manage and live independently should be carefully planned, adolescent friendly, and developmentally appropriate. Formats for this process might include books, educational handouts, Chronic Disease Self-Management Program workshops, peer mentors, health educators, web-based portals, mobile health apps, community health workers, and even music therapy.[40,41] The responsibility for decision-making should be increased gradually as the patient move towards

the adult care model. Institutions are piloting programmes that teach adolescents how to schedule an appointment with an adult provider, understanding medical insurance, and understanding medication prescriptions, and ordering refills through experiential learning.[44] An educational decision making programme could be introduced to those about to undergo transition to include both essential life and health-care skills as recommended by Calhoun and colleagues.[45] This work implemented an intervention with skill-based educational handouts based on areas of deficits with 61 adolescents to prepare them for the management of their disease as adulthood. The skills programme positively affected the participants' ability to manage their own care and their daily lives, including laundry, housekeeping skills, health-care skills, relationships, and sexuality.[45]

Increasing self-efficacy through a person-centred approach

Outcomes for adolescents with sickle cell disease moving to adult services are not always limited to perceived and actual prognostic markers of their illness, such as emergency department admissions and patient-reported pain scales, but also psychological and social factors, including anxieties about achieving future goals and self-esteem. Many young people find that although they might have the capability to manage their condition, they have low levels of motivation to do so and have developed behaviours that are just as likely to contribute to poorer prognostic outcomes.[30] Self-efficacy is defined by the psychologist Albert Bandura (Emeritus Professor, Social Science in Psychology, Stanford University, Stanford, CA, USA) in his social learning theory, as "one's personal belief in one's own ability to succeed in specific situations or accomplish a task".[46] A person's sense of self-efficacy plays a role in how they approach and handle tasks and challenges. A strong positive correlations between self-efficacy during paediatric to adult care transition and positive patient outcomes in patients with sickle cell disease is well established in the literature.[40,47,48] Sobota and colleagues,[47] constructed an expert panel of adult sickle cell health-care providers and asked them to complete surveys in an attempt to identify the most important indicators of a successful care transition. After surveying 78 health-care professionals, they found that the patient's self-efficacy was considered a top predictor of a successful transition. High self-efficacy in disease management is highly correlated with adherence to treatment, medication use, and outpatient attendance for young people undergoing care transition. The Sickle Cell Self-Efficacy Scale was the first tool used to test self-efficacy in adolescents and young adults with sickle cell disease, assessing how they responded to every-day activities.[49] Clay and Telfair[49] surveyed 172 patients aged between 11 years and 19 years, administering the Sickle Cell Self-Efficacy Scale in a cross-sectional study. Their findings mirrored those of a 12-month longitudinal study of adults with sickle cell disease by Edwards and colleagues,[50] which reported lower self-efficacy was associated with increased pain severity and increased psychological symptoms, and that improving self-efficacy led to improved symptomatic relief.

Self-efficacy is not static and can change depending on the situation and circumstances surrounding the patient; people can increase or lose their self-efficacy. This spectrum of self-efficacy is important: if low efficacy is identified early, it can be addressed well ahead of the transfer to adult care, to better prepare the young person. This is supported by the fact that those starting the transition process after 21 years of age are less likely to transition successfully when compared with patients who start the process under this age.[51] This might lend to the argument that without motivation or self-belief patients become more reliant on paediatric services, which are inherently more ordered. Care transition is a process not an event, and self-efficacy in particular must be ingrained from a young age. Self-efficacy can be improved by engaging individuals in the transition process long before the actual transfer of care[47] and encouraging person-centred planning at each health-care encounter to put the young person at the centre of decisions that affect them. When young people are meaningfully involved their attitude, behaviour, and learning can change and make them active partners who work with health-care providers to successfully manage their personal health ambitions. There might be times when the child or young person's stated goals seem unrealistic, but in a person-centred approach it is not for professionals to restrict a young person's dreams.[34,40,45] Rather, the professional's role is to think carefully about what the goal means to patient and help decide on concrete and realistic next steps that will move them in the right direction.[40,49–53] Ekman and colleagues[52] proposed that patient-centred care can be implemented in three stages. First, initiating any consultation by allowing the young person to tell their story (the patient narrative), which allows for the young person's views to be at the centre of their care. Second, developing the partnership through constant communication between the young person and health-care providers in which the young person's views guide any change or intervention. This is accomplished by sharing experiences and building a common understanding of the care plan. Third, safeguarding the partnership by documenting the decisions made and the preferences of the young person. The person-centred approach gives young people the opportunity to assume an active role in the decision making processes that directly affects them.[53]

Coordinating transition through community health workers or navigators

The importance of pretransfer and post-transfer care and support offered to young people and their parents and carers, who are often neglected in the transitional process,

cannot be emphasised enough. To facilitate successful care transitions, community health workers and patient advocates are frequently used to serve as advocates and educators to improve disease management, social support, and health-care navigation.[54] Community health workers are particularly useful in low resourced communities with a high disease burden. They are closely acquainted with the community they serve because they are often members of that community. The roles and responsibilities of community health workers can be tailored to unique needs and settings. For example, medication adherence is an important aspect of sickle cell disease management and transition readiness. The Hydroxyurea Adherence for Personal Best in Sickle Cell Treatment study,[44] a randomised, controlled, feasibility pilot trial aimed to increase hydroxyurea adherence by sending text message reminders to patients—a community health worker intervention. Improved adherence and health-related quality of life in youth with sickle cell disease was reported.[55] Although this study did not specifically intervene in health-care transition outcomes, such as completion of first adult provider visit, adherence and health-related quality of life, which have been highlighted as important transition indicators in literature, were assessed.[56]

A potential barrier to involving community health workers in improving the management of sickle cell disease is that some workers have a poor knowledge of sickle cell disease. One study which assessed the sickle cell disease-related knowledge of CHEWs in Nigeria found that only 69 (38%) of 182 health-care workers had a good knowledge about sickle cell disease and disease care.[57] Therefore, regular and comprehensive training is essential. Evaluation of the CHEWs programme is important to improve the quality of monitoring and care provided to individuals with sickle cell disease.[58] In addition to the CHEWs, transition navigators or coordinators have increasingly become involved in care transition programmes. Transition navigators provide disease and self-management knowledge and health-care system navigation. Unlike community health workers, transition navigators tend to be health-care professionals, such as social workers or nurses, and can be active members of both the paediatric and adult clinical teams.[31,59] Transition navigators responsibilities include reminding the patient to go to clinical visits and check-ups and helping the patient to adjust to more proactive management of pain.[60] In a structured transition programme with a transition navigator, there was a decreased number of transitioned youth lost to follow-up and improved or maintained medication adherence compared with youth who received standard care.[31] Several expert centres advocate for a psychologist to also be part of the dedicated transitioning team to help with the development of skills to manage stress and to support the development of a positive identity and self-efficacy.

Knowledge transfer through health promotion

WHO defines health promotion as "The process of enabling individuals to increase control over and improve their own health—a mediating strategy between people and their environments, synthesising personal choices and social responsibility in health".[61] An essential part of health promotion in young people with sickle cell disease is to ensure that they understand their diagnosis, the reasons for treatment, and have realistic expectations for their future. As such, care transition should aim to not only maintain their medical, physical, and psychological stability, but also prepare them for the future and the challenges that they might incur.

Sex and reproduction are extremely important to adolescents and young adults. However, how sickle cell disease affects these aspects of health is not widely taken into account and in some settings cultural and religious practises and beliefs potentially inhibit discussion. The unpredictable trajectory of sickle cell disease can make for difficult but nonetheless important conversations to ensure that young adults make informed choices regarding their future care and recognise the importance of ongoing engagement with health-care providers. Where resources are available, digital technologies, such as health information tools, have not been assessed, but are expected by adolescents to be potentially helpful.[35] Models for discussion, such as the Reproductive Life Plan[62,63] can be adapted for individuals with sickle cell disease. The cost-effectiveness of these different tools might vary according to the specificities of the countries and should to be evaluated accordingly.

Linking to adult services to ensure continuity of care

In a study using a Delphi analysis to assess indicators for a positive health-care transition, the timing of the first visit to the adult health-care centre, and trust between the adolescent with sickle cell disease and their health-care providers in the adult system were underlined as the two main factors.[47] Presenting the adult team (doctors, nurses, and psychologist) to young people through informational materials or (more preferably) during a visit to the adult centre, might decrease fears related to the transition. Alternating visits to the paediatric and adult centres and visits with both the paediatric and adult system physicians have been proposed.[59] Such strategies, that would likely not come at much additional cost, could increase patients' trust in the adult team.

Where resources are available, groups mixing adolescents and young adults might allow the older patients to help their younger counterparts to find their own answers and gain more insight into the disease by sharing their experiences. There have been many suggestions that in high prevalence areas the ideal scenario would be for the long-term care of young people with sickle cell disease to be provided by very

specialised multidisciplinary teams, with both paediatric and adult experience.[64] Adolescent health-care centres that bring together expert interdisciplinary care, excellent training for care providers, and cutting-edge adolescent health research might provide the leadership that can improve both the health of the adolescent population and increase their access to high-quality, holistic health services.[17]

Evaluating readiness for transfer of care

Many studies have evaluated the transitional period—the long-term process from adolescence through to adulthood—but disregard the transfer process, an isolated incident likely to have a large effect on transition. There is agreement between studies that transition readiness should be assessed before final transfer of care. Transition readiness is defined as the "adolescent's readiness to assume complete responsibility for their health care and their readiness to transfer to adult medical care".[65] The Social-Ecological Model of AYA (adolescent and young adult) Readiness for Transition (SMART) is a model used to identify components of transition readiness, including modifiable factors, such as the patient's knowledge about their disease, and non-modifiable factors, such as sociodemographics.[66] This model is generic and chronic diseases differ phenotypically and by the stigma they hold. Mulchan and colleagues[67] examined the applicability of the SMART model for those with sickle cell disease by interviewing a small sample of adolescents and young people who were undergoing or had recently undergone care transition and ten expert sickle cell disease health-care professionals to identify themes, which were evaluated to establish their applicability to the SMART model. They found that the SMART model might be an appropriate tool for assessing the readiness of patients with sickle cell disease for care transition, but factors such as stigma, race, and pain management were salient for those with sickle cell disease and should be taken into account when using a model of care transition readiness in patients with sickle cell disease. Porter and colleagues[68] investigated the perspectives of the care transition readiness of young adults with sickle cell disease who have transitioned from paediatric to adult health care. They conducted focus group discussions using the SMART components as a framework to interpret the findings. A major theme that emerged was developmental maturity. Some of the participants expressed that not everyone is prepared enough for care transfer at (or around) the age of 18 years and that there should be an age range in determining readiness.

Parental and guardian input is also important, and their involvement is often neglected when reviewing the literature regarding care transition. The Self-management and Transition to Adulthood with Rx (STARx) questionnaire and assessment is used as a measure of transition readiness of adolescents with chronic health-care conditions and is taken by the patient themselves. Nazareth and colleagues[69] used the newly adapted STARx-P a proxy questionnaire for parents and showed good internal reliability and consistency between the reports from the parents and the patients. Therefore, both youth and parental reports proxies are likely to be useful in understanding readiness of patients with communication difficulties, and for ensuring that appropriate measures are incorporated within the process based on need.

Saulsberry and colleagues[41] identified that attempts to improve the care transition process were limited by a small number of quality indicators to measure its success. Successful transition is often billed as the young person with sickle cell disease attending their first adult service appointment and using adult services for 1 year in the post-transition period.[31] However, this short time frame provides an incomplete picture and the focus on medical centre visits tends to neglect the majority of aspects that will also affect the individuals experiencing transition. Keeping medical appointments might not account for the readiness and motivation of a patient to engage in the process of a useful visit. A broader picture of health-care service use might be captured by recording the ratio of emergency care to scheduled ambulatory care visits or the medication possession ratio, but these ratios could reflect regional differences in health-care structure as much as the individual's ability to navigate the health-care system.[70] Other outcome measures include patient empowerment scales, transition readiness questionnaires, health behaviours, patient reported quality of life, parent-reported scales of their child's transition readiness and behaviours, and health-care usage.[18,31,40,71] Greater consistency of how success is measured would enable more comparisons to be drawn between different transition models.

Recommendations

Transition planning is complex and requires effective collaboration to maximise outcomes, especially in individuals with a chronic life-long illness. Poor transition affects health-care systems by increasing pressures and workloads (especially for nurses) and patients with worse transitional outcomes will tend to use emergency departments and acute care services more frequently because of poor medical adherence with preventive care. A globally standardised solution to care transition is unlikely to be practical given the substantial differences in cultural practices and barriers to care transition, such as resource limitations. Nonetheless, a shared goal is urgently needed with principles that could be adapted to meet the different local needs worldwide. Our recommendations for care transition planning goals for all children with sickle cell disease are outlined in panel 2, they are based on the SICKLE recommendations.

The systematic review by Campbell and colleagues[9] found that short term transition training workshops only have a small effect on the readiness for transition of young people with a chronic disorder. However, a more sustained approach, described by Hobart and Phan[17] defines paediatric to adult health-care transition as the process of transition that should be longitudinal and jointly undertaken by service providers and families with the young person at the centre. Educational objectives for health-care transition are to increase awareness of the disorder and strategies for self-management, adherence with medications, and the ability to navigate care services. We suggest that the benefits of adolescent transition programmes could be examined in the framework of a socioecological model of health, including health economics. Early data from the Sickle Cell World Assessment Survey[72] indicate that sickle cell disease has large indirect economic costs, including loss of hours of work every week, low educational attainment, and unfulfilled career potential. These economic costs could have enormous significance in LMICs where sickle cell disease has a high prevalence. We call for adolescent transition programmes to intentionally collect data to estimate the long-term effect of sickle cell disease on health outcomes, efficiency of health-care services, educational and career attainment, and other indirect economic costs. This data could help to describe the economic returns of investment into resources devoted to adolescent care transition programmes.

Implications of current patterns and future directions

In HICs care transition programmes have been created to prepare adolescents and young adults for adult-centred care by improving their disease knowledge and self-management skills and assisting them with the navigation of the adult health-care system. Despite this, few programmes have been formally evaluated and a evidence-based care delivery model for sickle cell disease has not been reported. As stated earlier, there is no consensus on the definition of successful transition, with success being measured by narrow and restricted outcomes, such as attendance to first provider visit and hospital use. More research is needed to standardise comprehensive health-care transition outcome measures that assess outcomes longitudinally and test care delivery models.[34,41] Outcomes need to be expanded to include care cost, access, and gaps in care coordination; additionally, patient outcomes, such as quality of life, satisfaction with care, treatment adherence, cognitive functioning, and academic and vocational attainment. Furthermore, collaboration across paediatric and adult health-care systems and information gathered from registries and large data sets across institutions are essential to provide a complete picture of the transition process. Sustaining care transition programmes can be a challenge, given the

***Panel 2*: The SICKLE recommendations**

Skills transfer

Patients should be empowered from an early age to manage their own condition by teaching them the necessary skills as soon as developmentally appropriate, including how to self-manage symptoms, such as pain; how to recognise when professional advice is required; and how to book their own health-care appointments.

Increasing self-efficacy

Every young person's voice should be at the centre of their health-care plan and their beliefs, goals and motivations should be discussed at each encounter to ensure they are active partners in managing their care. Parents and caregivers are important support systems for young people with sickle cell disease. Their knowledge and anxieties about the care transition process should be addressed so they can optimally support the young person's development of self-efficacy.

Coordination of transition

Every young person transitioning from paediatric to adult care should be supported by a named community health worker or nurse navigator who is appropriately trained in providing guidance on disease knowledge, self-management skills, medication adherence, and health-care system navigation. Paediatric and adult providers must maintain communication to ensure optimal transfer of care.

In low-income and middle-income countries, community health extension workers, clinical medical officers, and village council mobilisers should be trained and retrained specifically to implement and aid transition by offering continuous education, working with patients and their families to achieve self-efficacy and organise the various linkages to specialist care and transfer of sickle cell care to adult services. They already assist in education of patients living with other chronic diseases and are best suited to also reduce loss to follow up.

Knowledge transfer

All patients should have a good understanding of sickle cell disease, including its cause, signs and symptoms, potential complications, management, risks of non-adherence to medication (hydroxyurea and penicillin), and prognosis before transfer of care.
We recommend that multidisciplinary (including young people with sickle cell disease and their families) task forces develop a curriculum and handbook for the transition from paediatric to adult care.

Linking to adult services

All young people should know when their care will be transferred and who the adult provider is or could be, and they should ideally have either a joint first consultation or a period of overlap whereby their first consultation with their adult care provider occurs before their last paediatric appointment. They should also be introduced to other services that they would need to access in the course of their adult care.

We recommend a mapping of all facilities offering specialist sickle cell care in lower-middle income countries for easy referral and identification of the nearest adult centred service for patients in care at paediatrics only centres.

Joint paediatric and adult care clinics should be established where available resources permit. A minimum requirement is to offer such a clinic before care transfer, which might be as early as 12 years for adolescents who transfered at younger ages to adult oriented care, as done in the Middle East and countries in sub-Saharan Africa. Such an initiative is best driven by nurses and trained community health extension workers, engaging both paediatric and adult haematologists in multidisciplinary care.

Evaluating readiness

Before transfer of care, transition readiness should be assessed to ensure every young person is developmentally ready to assume complete responsibility for their healthcare and adequately prepared for doing so. Tracking the progress of the young adult for several years will enhance evaluation of the success of adolescent transition programme.

Search strategy and selection criteria

We searched PubMed, Ovid, and the Web of Knowledge, using the search terms "sickle cell disease" and "transition" from inception of the databases to Aug 31, 2019, and also searched the references from relevant articles identified. Only peer reviewed papers that were available in English were included and relevant studies cited in the recovered works were also used for analysis. Reports from meetings were included only when they related directly to previously published work. We did not limit the years of publication because some were used for historical references to paint the picture of how practice has changed over time. Experience from the global taskforce assembled to create this Review was also drawn upon and local and national guidelines were included where applicable, thus providing a perspective of global practice.

pay structure for health-care services in some HICs; therefore, developing alternative reimbursement models is necessary.[30] Future research into care transition interventions should make use of comparative effectiveness studies to determine which health-care interventions provide the most benefit to patients and allow for the dissemination and implementation of science approaches to inform how to best adopt, implement, and maintain transition interventions.[34,41] Finally, we need to continue to include the voice of patients through qualitative research and administration of patient-reported outcomes measures to ensure care transition needs are identified and addressed adequately.

In LMICs, similar to the HICs, the goal for transition is to prepare young people with a chronic illness for adult health-care services using the six SICKLE recommendations. However, implementation is hampered by prevailing persistent shortages of person power and health-care personnel trained in sickle cell care. Furthermore, there are many standalone paediatric sickle cell disease clinics with little or no existing relationships with adult-centred care. Barriers to achieving the desired seamless transfer from paediatrics to adult oriented care provisions are substantial. In some cases, transfer is done by letter or, as reported in some centres, the clinic administrator automatically moves the patient to the adult ward based on the patient's date of birth. The over reliance on paper records and file systems, which in some cases are handed over to patients to carry to the adult providers, are some of the challenges that could be addressed through policy changes with little funding implications. Furthermore, health funding and poor health insurance coverage mean that the cost of care and clinic attendance are often borne by the family through out of pocket expenses. The consequence is that clinic attendance and medication adherence are effected in some scenarios by costs rather than by motivation and behavioural skill. Youth education before transfer to adult centres could be supplemented by the use of digital solutions and social media platforms, as reported by Creary and colleagues.[69] The proportion of youth exposure to information technology and mobile phones is increasing, even in LMICs. Digital solutions also offer the opportunity to create awareness of sickle cell disease, disease complication, and treatment options in health-care providers and equip them with a directory of specialist services for patient referral.

Structured care transition planning for LMICs is needed to enhance the content of information and records that are shared with adult centres, including medical results and treatment history in a succinct way. It is time to undertake further research to describe the transition related activities, the health-care delivery system barriers, and health-care funding models in LMICs in detail. The ARISE initiative provides the platform for mentoring partnerships between specialist health services with robust care transition services and services in LMICs, which could help the development of good care transition practices in the latter setting. Peer mentoring for affected youth and younger patients could also empower the adolescents to better embrace the transition to adult-centred care.

The prevalence of digital technology in both LMICs and HICs is on the increase, this is more so for young people. In addition to accessing websites with health-related educational material, youth also look to social media platforms and online communities to learn more about various disease-states or to obtain answers about personal health concerns.[73] Although access to digital technology is variable between youths in LMICs, 40% of children in Africa aged 15–24 years were estimated to be online in 2017 91·9% more than 5 min per day, 69·4% more than 30 min per day, and 36·6% more than 1 hr per day for health-related internet use.[74] Altogether, this increased internet usage suggests that efforts to engage adolescents with sickle cell disease at the time of care transition through social media and other web-based technologies might be a crucial mode for more widespread implementation of transition best-practices. Creary and colleagues[69] reported the use of mobile health strategies in adolescents for symptom tracking, communication, and to deliver treatment. Using implementation science to test mobile health technologies will enable proper evaluation and adaptation that accounts for context (where patients and providers are), barriers affecting all stakeholders, and the perceptions and preferences of users (patients and care providers).[74]

Conclusions

Successful transition from paediatric to adult services for adolescents and young adults with sickle cell disease is fundamental in the treatment and management of their condition and essential to improvements in their

survival, both quantitatively and qualitatively. The six SICKLE recommendations, highlighted in this Review, that should be considered in any sickle cell disease care transition plan for young people are skills transfer, increasing self-efficacy, coordination of transition, knowledge transfer, linking to adult services, and evaluating readiness.

Objective goals and outcome measures must be established on a global scale to ensure maximum benefits of transition programmes and enable cross programme comparisons. Multisite longitudinal, prospective studies are urgently needed to see what more can be done to address this growing public health need.

Contributors

BPDI and MT supervised the development of the Review. BPDI, SMC produced the original concept draft. CES review the original draft and designed concept of benchmarking strategies and recommendations. SMC, JP, LLH, WA, IDA, and NA review the literature. LLH contributed to the additional global concepts. IDA produced the sub-Saharan African perspectives. NA produced the global perspectives. All authors contributed to the writing and review of the submission.

Declaration of interests

BPDI reports grants and personal fees from Novartis, AstraZeneca, Cyclerion, and Global Therapeutics; and grants from Pfizer, Nova Labs, Celgene, and Add Medica, outside the submitted work. LL-YH reports personal fees from Aruvant, Emmaus, and Pfizer; grants and personal fees from Astra Zeneca, Cyclerion, Forma Therapeutics, Global Blood Therapeutics, Novartis; and grants from Hilton Publishing, and Imara, outside the submitted work. MDM reports grants and personal fees from Novartis; grant, personal fees and non-financial support from Addmedica; and grants from bluebird bio, outside the submitted work. BA reports personal fees from Imara, bluebird bio, Cyclerion, NovoNordisk, Terumo BCT, Novartis, and Vertex, outside the submitted work. MRA reports grants and personal fees from Novartis; and grants from Vertex pharmaceuticals and AstraZeneca, outside the submitted work. MT reports personal fees from UptoDateR, outside the submitted work. All other authors declare no competing interests.

Acknowledgments

ARISE has received funding from the European Union's Horizon 2020 research and innovation programme under the Marie Sklowdowska-Curie grant agreement No 824021. National Heart Lung Blood Institute K01 HL125495 Identification of Predictors and Outcomes of Sickle Cell Health Care Transition. We thank Leon Tshilolo, Priyanka Samal, Hatoon Ezzat, and Jane Hankins for personal communications and invaluable advice on international transition cohorts

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