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Samir K. Ballas
Thomas Jefferson University

Gaye Riddick-Burden
Thomas Jefferson University

Elisabeth Congdon-Martin
Thomas Jefferson University

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Samir K. Ballas[1], Gaye Riddick-Burden[2], Elisabeth Congdon-Martin[3]

[1]Cardeza Foundation for Hematologic Research, Department of Medicine, Sidney Medical, Thomas Jefferson University, Philadelphia, PA

[2]Department of Medicine, Division of Internal Medicine, Thomas Jefferson University, Philadelphia, PA

[3]Department of Medicine, Division of Hematology Thomas Jefferson University, Philadelphia, PA

Corresponding author:

Samir K. Ballas, Cardeza Foundation for Hematologic Research, Department of Medicine, Sidney Kimmel Medical College, Thomas Jefferson University,
1020 Locust St, Suite390, Philadelphia, PA19107
Phone 856-745-6380
Fax 856 7950809
e-mail: samir.ballas@jefferson.edu

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Abstract

Transition of adolescents with sickle cell disease (SCD) to adult programs is associated with increased morbidity and mortality. The reasons for this poor outcome are not well known. This report describes the various factors that affect the outcome of the transition process. These include four inter-personal factors: country of residence, region within the country of residence, the health care system and intra-personal factors. Each factor is described in some detail. Understanding these factors and the establishment of guidelines or recommendations could improve the outcome of this critical transition in the life of patients with SCD.

Introduction

Transitions are inevitable facts of life. Differences between an old and a new phase of life could be a stark landscape that threatens the sense of security. We all experience many life transitions. With time, most of us acclimatize to the new change and achieve a new sense of stability and security. This sequence of events, however, does not occur when most children and young adults with sickle cell disease (SCD) transition from pediatric to adult programs. Besides the usual personal transitions, they face an additional serious transition of their medical care to adult programs. Transition of medical care includes changes to the level, place or providers of medical care as patients and families negotiate the complex health care system [1]. This process is lengthy, tedious, expensive and its outcomes are often unpredictable, necessitating circuitous trials. This transition is associated with increased morbidity and mortality as was described previously [2]. There is a treacherous deep gap (Figure 1 Panel A [3]) between the pediatric and

adult programs and most children are not aware of it. The ultimate results of this transition are dependent on several factors as shown in Figure 1 Panel B as well as personal experience and publications by patients treated in different sickle cell centers across the United States [4-13]. This commentary reviews the determinants of the outcome of this transition.

The best method to determine the difficulties experienced by patients with SCD after transition to adult care is to quote how the patients describe their own experience. To that end, there are at least 9 books written by adult patients with SCD in which they describe the various issues they encountered [4-12]. These will be referred to as needed in this commentary.

1. Inter-personal conflicts

a. Conflicts with adult providers

Medical care of children and adolescents with SCD is usually provided by pediatric hematologists within a framework of state-of-the-art care including all aspects of the disease. Physicians, nursing staff and ancillary personnel in the hospital and clinic demonstrate empathy and provide comprehensive multi- disciplinary care.

The excellent care provided by pediatricians is eclipsed when the children are transitioned to adult programs. Lack of empathy and mistrust complicate matters. Young adults with SCD are surprised to have limited access to hematology specialists or other providers specialized in caring for patients with SCD. Accordingly, they try to go back to their pediatricians, seek care in any emergency department (ED), wander along a maze of maladaptive

behavior. As a result, some young adults continue to be seen in pediatric clinics and EDs until they are in their late twenties or early thirties [14].

b. Conflicts with Emergency Departments' Personnel

Emergency Departments (EDs) and their personnel are often the nemesis of patients, especially adults, with SCD. One patient referred to the ED as "hell" and other patients referred to it as "asylum" [4,6]. Emergency departments are the last resort patients go to after failing to control their pain at home. The patients expect waiting for long periods before they are screened and longer before receiving the desired treatment, if any [15]. Most painful is the body and verbal language of some ED personnel including humiliating sneers and jeers [16]. Unfortunately, some patients may not seek care at all until they end up in an emergency room with potentially life-threatening complications. In our experience, a few patients turned away from the ED, were found dead at home the next day.

c. Conflicts with parents or guardians

Rarely, some parents/guardians may declare that they are not responsible to take care of their children once they are legally adults aged 18 years or more. In these cases, the young adults have no choice but to pursue undesirable behaviors. More often, however, it is the young adults who declare that they are adults now who can make their own decisions and not follow advice from parents or guardians.

d. Conflicts with pediatric providers

Although pediatricians provide excellent care for their patients with SCD, rarely, they create a conflict in certain situations. Occasionally, a teenager less than 18 years old is considered a problem patient because he/she has frequent painful crises (VOCs) that require

frequent and lengthy hospital admissions and the use of high doses of opioids. Such behavior is considered typical of adults with SCD and, hence, justifies the transition of such patients to adult programs. Most adult programs, however, refuse to accept these patients but a few do. The germane issue in such situations is that the patient in question will be labeled a problem patient that negatively affects his/her management as an adult for a long time.

Another situation pertains to pregnant girls. These are logically transitioned to obstetrics and considered "adults" whose medical care should be provided by obstetricians, adult hematologists or providers. Pediatricians consider a pregnant girl an adult and adult hematologist consider her a child since she is less than 18 years old. There are no clear guidelines what to do in such situations.

Another emerging barrier to the use of opioids in the management of sickle cell pain pertains to the attitude of pediatric residents. Fearon et al [17] reported that certain negative views of patients with SCD were prevalent among all pediatric residents. Moreover, less experienced residents were less comfortable treating acute sickle cell pain and were more concerned with addiction compared with more experienced residents.

2. Country of Residence

Population-based studies in the United States (US) showed a significantly higher rate of deaths due to SCD in young adults compared to children or teenagers [18,19]. The transition from pediatric to adult care is associated with loss of comprehensive care and seems to be the major risk factor. Moreover, poor socio-economic conditions, different cultural and educational factors probably also play a role in the outcome of transition. Recent study by Le et al [20] did

not confirm the dramatic increase in SCD-related mortality among young adults in Belgium as was observed in the US [18,19]. The Belgian national healthcare system covers the medical costs irrespective of patient age and employment. In addition, in Belgium there is a strong network of hematologists that shares standard of care and patients have direct access to specialists and secondary or tertiary hospitals [20]. Moreover, evictions, disparities, faulty accusations, rationing of care, etc. are not major issues in Belgium the way they are in the US [8-10]. Noteworthy, the outcome of transition in Belgium is impressive despite the fact that most patients with SCD in Belgium are immigrant from central Africa a region known to have more severe forms clinical SCD [20].

3. Geography/Region within the country of residence

Specific geographic issues within the country of residence should be considered in analyzing the factors associated with increased morbidity and mortality after the transition of adolescents and young adults with SCD to adult care. The Multicenter Study of Hydroxyurea (MSH) in sickle cell anemia showed that management of VOCs at home, in acute care facilities, and in the hospital, seems to be sex, age, and geographic region dependent [21]. Specifically, the choice of the route of opioid administration was region dependent with the frequency of utilization of oral opioids at home was significantly highest in the Northeast and lowest in the West and the frequency of utilization of parenteral opioids in acute care facilities tended to be highest in the West and lowest in the Midwest.

Anderson et al [22] reported that during 10 years of follow-up after transition from pediatric to adult care, the rate of death in Atlanta was much lower than that in Philadelphia: 5.8% in Atlanta versus 22.2% in Philadelphia.

4. Health care system

The health care system in the US is the Achilles heel of the transition from pediatrics to adult care programs. Its weakness is the major cause of disruption of the quality of care in the adult setup [23]. The frequency of morbidity and mortality after transition to adult programs is associated with the health insurance the patients have. Patients who had commercial coverage had significantly less VOCs, hospital admissions and mortality than patients covered by Medicaid [24]. Thus, the health care system is the major determinant of the outcome of transition from pediatrics to adult care.

5. Intra-personal Factors

In post-adolescence, young adults with SCD feel free from adult interference. They think they are now autonomous, having the right of self-determination. As a result, adherence to medical care, follow-up, medication schedules, etc. gradually fade away. This happened in the case of children who were on chronic blood exchange transfusion due to ischemic stroke. Once they became adults aged 18 years, they refused to continue having exchange transfusion on a regular basis as before despite all advice by their parents, pediatricians and adult hematologists. These patients died within 3 -5 years after transition due to complications of their disease, mostly recurrent severe stroke [25].

In Brazil, health coverage is not a major issue for patients with SCD. Patients who have no private coverage are covered by the State. Patients are usually assigned to a Medical facility close to their residence. Children followed in such Medical facilities continue to be followed in the same facility when their care is transitioned from pediatrics to adult programs, after the age of 18 years. In spite of this, morbidity and mortality increased significantly after transition as shown in Figure 2 [26]. Lack of regular follow ups, compliance to treatment, family support and lifestyle were the important factors contributing to the increased morbidity and mortality as described at a single institution in Rio de Janeiro [26].

Conclusion

Understanding the determinants of transition outcome allows the establishments or the revision of guidelines and recommendations for the transition process. Most important among these is that no child below the age of 18 years should be transitioned to adult care due to frequent VOCs that require relatively high doses of opioids. Similarly, pregnant girls should be transferred to Obstetrics care during the pregnancy/post-partum time and back to pediatrics after that. The American Academy of Pediatrics reviewed current trends and issues related to adolescent pregnancy including the legal and policy implications of concern to pediatricians [27]. On the other hand, transition to adult programs should not be delayed beyond the age of 25 years in the US in order to maintain coverage by the parents' health care status.

Once pediatricians realize that transition to adult programs should usually be between the ages of 18 -25 years, the transition of each specific patient becomes individualized. Howard et al [28] established a transition clinic for that purpose. St. Jude Hospital in Memphis MO also has

transition clinic for patients with SCD. In such a clinic the situation of each patient will be analyzed within the framework of his/her psychosocial status, family structure, education, severity of SCD and potential for employment. The clinic will include a pediatrician, adult provider specialized in SCD or a hematologist, nurse practitioner and a social worker. Health care coverage is often a problem in patients covered by Medicare or Medicaid. Benefits applied to patients with SCD vary among states. It is best if the billing department of the Institution in question helps finding the approved coverage for each patient according to the Affordable Health Care Act and beyond if applicable. The rules are complex and are described by the Centers of Medicare & Medicaid Services [29].

Legends to Figures:

Figure 1: (A) The treacherous gap in challenging transitions. Adapted from Science 2019;363:24-26 with permission. (B) Determinants of the outcome of transition of children with sickle cell disease to adult programs.

Figure 2: The mortality rate in under 5-year-old patients was lower than the mortality rate in the 6–11 age group. However, there was a significant difference in the mortality rates between the 6–11 and 12–18 age groups. After the age of 18 years, there was a significant and sudden increase in the mortality in the 19–29 age group. The mortality continued to increase significantly after the age of 30 years. From Rev Bras Hematol Hemoter 2018; 40(1): 37-42 with permission.

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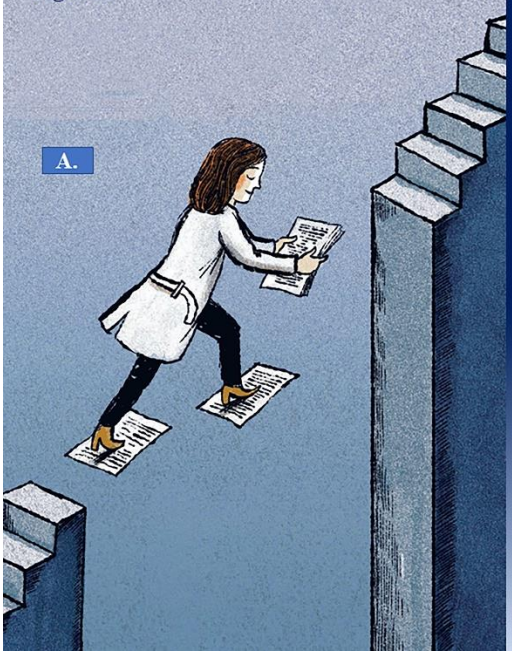
Figure 1



B.

Determinants of the outcome of transition of children with sickle cell disease to adult programs:

1. Inter-personal conflicts
    a. Conflicts with adult providers
    b. Conflicts with Emergency Departments' Personnel
    c. Conflict with parents or guardians
    d. Conflicts with pediatric providers
2. The country of residence
3. Geography, region and weather within a country
4. Health care system
5. Intra-personal factors

Figure 2