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Beyond the transition of adolescents and young adults with sickle cell disease to adult care: Role of geography.

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Beyond the transition of adolescents and young adults with sickle cell disease to adult care: Role of geography

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The transition of medical care of patients with sickle cell disease (SCD) from pediatric to adult providers represents a milestone in their lives. Major concerns among adolescents and young adults about transition include taking responsibility for self, making own decisions, cost of medical care, fear of suboptimal pain management, and reluctance to leave known providers. Survival among children with SCD increased progressively over the last 40 years. The advent of newborn screening, antibiotic prophylaxis, newer vaccines, safer blood transfusion, and hydroxyurea are the major factors that prolonged the survival of children with SCD. Consequently, over 95% of children with SCD live to be adults in the Western world (Europe and North America) and other countries especially in Jamaica and Brazil [1-3].

Unfortunately, despite this impressive improvement in survival among children and adolescents,, it suffers from an eclipse with a setback to increased mortality after the transition of medical care from pediatrics to adult medical care [4]. Thus, a high risk of death is associated with the time during and shortly after the transition to adult medical programs [5]. Hamideh and Alvarez [6] reported that SCD mortality in the US in 1999 – 2009 compared to 1979–1998, significantly decreased by 61% in infants <1 year of age, by 67% in children aged1–4years, and by22–35% in children aged 5–19 years. After 19 years of age, mortality rates increased from 0.6 in the 15–19 year group to 1.4/100,000 in the 20–24 year group. A recent study from California and Georgia also documented that the mortality rate increases after age 15 and this difference persists through adulthood [7].

The reasons that cause this surge in mortality after transition are not well known. Initially they were thought to be due to difficulties in identifying adult sickle cell providers or adult hematologists to enroll transitioned patients in their programs for regular follow-up and management. Although this is an important issue in the saga of transition, it is not the only reason.

The problems associated with transition are much more complex than thought before and are due to multiple factors including, but not limited to, poor or inadequate communication among patients, parents and providers, inadequate transfer of care from the pediatric team to the accepting future adult programs, negative experiences in the emergency rooms, access to hydroxyurea therapy, the country of residence, etc. [8, 9]. In this correspondence, we wish to emphasize the role of geographical variations in the same country that affect the outcome of transition. The geographical regions we will focus on include Philadelphia, PA [10] and Atlanta, GA [11].

Table 1 lists the major differences between the two cities. In Philadelphia 90 adolescents and young adults with SCD were followed prospectively for 10 years (1994-2004) after transition from the pediatric sickle cell program at St. Christopher Hospital to the adult sickle cell center at Thomas Jefferson University Hospital. Sixty-five of these patients had sickle cell anemia (SS), 16 had Hb SC disease and 9 had sickle-β^+-Thalassemia (S- β^+-thal). Their age at transition varied from 18 to 25 years. Twenty (22.2%) patients died within 10 years after transition and age at death was 24.9 ± 2.95 years (range 23-26 years). The rate of death was highest in patients with SS where 17 out of 65 patients (26%) died within the 10 years after transition. Complications of SCD after transition included stroke, acute chest syndrome, avascular necrosis, leg ulcers, anxiety, depression, priapism and multi-organ failure. About one third of the patients developed persistent pain between vaso-occlusive crises. Nineteen patients (10 males, 9 females) were employed.

The demographic of the patients in Atlanta are shown in Table 1. Most important among these is that the rate of death in Atlanta was much lower than that in Philadelphia: 5.8% in Atlanta versus

22.2% in Philadelphia during the 10 years after transition. The major cause of death was iron overload due chronic organ damage. It is most likely that these patients had chronic organ damage due to SCD that required frequent blood transfusions leading to iron overload.

The reason mortality after transition is lower in Atlanta than in Philadelphia are not known. The care systems available to the youth in the 2 cities differ because the Georgia Comprehensive Sickle Center at Grady provides emergency care 24 hours a day 7 days a week and has had an active transition program since 1985. Other possible causes include the warmer weather in Atlanta, the cultural and psychosocial community fabric in Atlanta may be more attentive to the problems associated with SCD and the genetic types of SCD in Atlanta may include those that are known to be associated with milder disease. These types include the Senegalese haplotype that is more common in Southern USA. In the Philadelphia area the Benin haplotype, known to be associated with more severe disease, is most common. Notably, The Multicenter Study of Hydroxyurea (MSH) in SS [12] showed that the number of crises was highest in the Northeast and lowest in the South and the average duration of a painful crisis was also highest in the Northeast but lowest in the West. The study of deaths in California and Georgia did not show any significant differences in death rate is the 2 state [7].

In summary, in analyzing the factors associated with increased morbidity and mortality after the transition of adolescents and young adults with SCD to adult care, specific geographic issues should be taken into consideration. The Multicenter Study of Hydroxyurea (MSH) in SS [12] showed that management of acute sickle cell painful episodes at home, in acute care facilities, and in the hospital, seems to be sex, age, and geographic region dependent. 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 was highest in the West and lowest in the Midwest. Geographic variations are emerging as important factors in analyzing results of randomized controlled trials [13]. These variations are specifically important in trials that include rare diseases that require enrolling patients from many countries across the globe.

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