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The Painful Face of Poverty

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On the 0 to 5 pain faces scale,1 the health care system for children with sickle cell disease (SCD) gets an excruciating 5. In a country where advances in care and research for SCD are praised as among the best in the world, children with this condition continue to receive inadequate health care and consequently fail to fully benefit from substantial progress in scientific research. In the United States, the disease affects close to 100,000 persons, most of whom are members of minority populations who suffer from poverty and sub-optimal access to adequate medical care. An alarming number of children with SCD are uninsured or do not receive care at specialized centers for SCD (Comprehensive Sickle Cell Centers). Moreover, the number of hematologists and other health care providers committed to the care of patients with SCD is insufficient to provide adequate care to this population. These problems are magnified after teenagers with SCD transition to adult care and not infrequently become dependent on emergency department services as their primary source of medical care.

Decreased access to adequate health care reflects another problem: poor compensation for the expensive cost of caring for SCD patients. As with any chronic pediatric condition, management of SCD requires more frequent and more substantial use of medical resources, but managed care organizations and pediatricians do not receive compensation rates reflective of the high cost of caring for these children.2

In the current issue of *Pediatric Blood & Cancer*, Raphael et al. have described health care utilization among low income children with SCD, compared to the general pediatric population. Their study examined the population size, demographic characteristics, continuity of insurance coverage, utilization patterns, and expenditures for children with SCD in a managed care plan exclusively designed for Medicaid and State Children's Health Insurance Plan (SCHIP) recipients in Houston. A remarkable number of pediatric patients with SCD were not retained in the Texas Children's Health Plan (TCHP) during the study period and therefore lost coverage, in marked contrast to the general TCHP population. The consequent gaps in health coverage are particularly significant for this vulnerable group of children, who often experience life-threatening events without advance notice. Predictably, the authors found higher emergency room, home health, and behavioral health utilization compared to the general population. Compounding the problem of soaring emergency care use was the sub-optimal fraction of children followed by a primary care provider (63%), which may indicate a trend toward utilizing the emergency department as the primary source of care. Taken together, these findings reflect the short reach of a health care system that fails to provide comprehensive, specialized, and continuous medical care to these at-risk individuals, who often experience the long-term effects of health care disparities.

A striking finding from the work by Raphael et al. is the very small proportion of children with SCD that are followed by a hematologist: a mere 10%. The opportunities that may be

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missed by pediatric patients with SCD who are not followed by a hematologist at a sickle cell center include: prevention of long term disease complications (e.g., monitoring for and treatment of end organ damage), prevention of stroke through TCD screening, education about SCD, academic support, preventive health maintenance and anticipatory guidance, early treatment of life-threatening complications, provision of immunizations, optimal utilization of hydroxyurea therapy, and participation in research. In fact, reduction of morbidity and mortality from SCD has been linked to care at a specialized center for SCD. 3

This study had inevitable limitations because only cross-sectional data derived from a large public database of claims information was utilized. These data suggested a progressive loss of insurance coverage as children aged, but this could only be implied as no longitudinal data was provided. In addition, claims data as the sole source of information is potentially compromised by incomplete or misclassified records. But in certain circumstances, the use of claims data is as good as it gets when analyzing large numbers of patients who are cared for by different providers at different institutions. Taking into account these limitations, the study is still very informative and eye-opening.

The road from efficacy to effectiveness in medicine is long and arduous, as progress may come slowly and changes are often difficult to be assimilated. Two examples of efficacious interventions that could improve health care for persons with SCD are hydroxyurea therapy and Transcranial Doppler ultrasound (TCD) screening. Despite having been shown to reduce painful events and hospitalization,4 hydroxyurea remains under-utilized, as demonstrated by the unchanged hospitalization rates in a population eligible for this treatment.5 Recently, a national consensus conference lead by the NIH also concluded that hydroxyurea is under-utilized.6 Transcranial Doppler ultrasound screening can identify children with SCD at risk for stroke,7 but many barriers hamper its widespread use and only a small fraction of appropriate subjects are currently benefiting from this technology.8

Research in SCD is currently in a period of major reorganization, based in part on recommendations from conferences for the following research priorities: 1) develop and expand curative therapies such as hematopoietic stem cell transplantation and gene therapy; (2) elucidate the genetic modifiers of SCD and integrate genetic information with biomarker, clinical, and psychosocial data; (3) expand and integrate basic knowledge of hematopoiesis and gene regulation, sickle red cell pathophysiology, vascular biology, inflammatory mechanisms, and coagulation regulation to refine the systems – biology model of SCD; (4) develop evidence-based treatment strategies based on improved understanding of pathophysiological mechanisms, genetic modifiers, biomarkers and environmental factors; and (5) translate research advances into clinical practice by developing and implementing evidence-based guidelines for medical and psychosocial SCD treatment and disseminating these therapeutic advances effectively. Recently, the NHLBI has indicated its “recommitment to SCD research”,9 indicating that it will reconfigure the Comprehensive Sickle Cell Centers into a Basic and Translational Research Program and a new Clinical Trials Research Network (CTRN), with the goal of gaining participation in clinical research to as many eligible investigators and individuals with SCD as possible.

In summary, the combination of a disease that combines progressive organ damage and interspersed with serious acute events, poverty, racial inequalities, disease stigmatization, uninsured or unstably insured children, and poorly compensated health care professionals results in a precarious situation. Studies like that of Raphael et al., are important to draw attention to problems in health care delivery as this information may help spur changes that will result in increased effort and resources, with the goal of a less painful future.

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