HHS Public Access

Author manuscript

Am J Prev Med. Author manuscript; available in PMC 2015 June 04.

Published in final edited form as:

Am J Prev Med. 2012 June ; 42(6): e101–e103. doi:10.1016/j.amepre.2012.03.005.

The Jamaican Historical Experience of the Impact of Educational Interventions on Sickle Cell Disease Child Mortality

Scott D. Grosse, PhD,
Division of Blood Disorders National Center on Birth Defects and Developmental Disabilities CDC Atlanta, Georgia.

Hani K. Atrash, MD, MPH,
Division of Blood Disorders National Center on Birth Defects and Developmental Disabilities CDC Atlanta, Georgia.

Isaac Odame, MB, ChB, MRCP,
Division of Haematology/Oncology Hospital for Sick Children Department of Paediatrics, Faculty of Medicine University of Toronto Toronto, Canada

Djesika Amendah, PhD,
African Population and Health Research Center Nairobi, Kenya

Frédéric B. Piel, PhD, and
Spatial Ecology and Epidemiology Group Department of Zoology University of Oxford Oxford, UK

Thomas N. Williams, PhD
KEMRI Centre for Geographic Medicine Research (Coast) Kilifı, Kenya

INDEPTH Network of Demographic Surveillance Sites Accra, Ghana

Department of Pediatrics and Nuffıeld Department of Medicine University of Oxford Oxford, UK

We welcome the opportunity to provide additional support for the assertion in our paper on sickle cell disease (SCD) in Africa that newborn screening can result in substantially reduced mortality even in the absence of specifıc interventions such as penicillin prophylaxis or pneumococcal conjugate vaccines (PCVs).[1] Three references were cited describing reductions in SCD mortality that occurred prior to the availability of those interventions.[2–4] In one U.S. study, the rate of mortality among infants with sickle cell anemia diagnosed as newborns in a pilot screening study during 1975–1985 was 1.8% compared with 8% among infants diagnosed after age 3 months, a difference attributed to better follow-up and education.[2] The subsequent introduction of oral penicillin prophylaxis in the U.S. was associated with a 36% reduction in SCD mortality in children aged <4 years between 1983–1986 and 1987–1990.[5] The administration of PCV-7 to U.S. infants

sgrosse@cdc.gov.

Newborn screening for SCD presents an important opportunity to reduce the burden of mortality in children aged <5 years in sub-Saharan Africa.

The fındings and conclusions in this report are those of the authors and do not necessarily represent the offıcial position of the CDC.

No fınancial disclosures were reported by the authors of this letter.

beginning in 2000, which eliminated many deaths from invasive pneumococcal disease (IPD) due to incomplete adherence to penicillin prophylaxis, was accompanied by a further 42% drop in SCD mortality in children aged <4 years between 1995–1998 and 1999–2002.[5]

In Jamaica, King et al. and others documented sharp reductions in SCD mortality among infants born between 1973 and 1981 who were enrolled in the Jamaican Sickle Cell Cohort Study (JSCCS).[4,6] The rate of death in the first 2 years of life fell by 71% from 14% in the 1973–1975 birth cohort to 4% in the 1979–1981 birth cohort. One specific intervention, parental education on the detection of enlarged spleens and need for medical attention, was shown to result in a 74% decrease in the death rate from acute splenic sequestration (ASS) from 1.3 per 100 patient-years during 1974–1978 to 0.3 per 100 patient-years during 1979–1983, calculated from data on incidence and case-fatality of ASS.[4,7]

Among the 1995–2006 birth cohort with SCD, the rate of death before age 2 years fell by a further 75%, to 1%, as a result of early-life interventions introduced after 1981.[4] Chief among these is penicillin prophylaxis, which in Jamaica involves monthly intramuscular administration of long-acting penicillin with ~90% compliance.8 Although pneumococcal polysaccharide vaccine has been administered to children aged 4 years in Jamaica since 1988, because of its cost few Jamaican children with SCD receive PCV.[9] The Jamaican sickle cell disease program is highly effective and can serve as a model for resource-limited settings with good access to health care. One sentence in the current authors' paper was interpreted to refer to contemporary SCD care in Jamaica, which was not the intent. King et al.[4] was cited as an accessible source of information on the Jamaican historical experience.[6,7] That experience demonstrates that newborn screening for SCD can result in dramatic improvements in child survival even if children are not provided with either penicillin prophylaxis or PCV, although both interventions are certainly important.

References

1. Grosse SD, Odame I, Atrash HK, Amendah DD, Piel FB, Williams TN. Sickle cell disease in Africa: a neglected cause of early childhood mortality. Am J Prev Med. 2011; 41(6S4):S319–S323. [PubMed: 22099353]
2. Powars D, Overturf G, Weiss J, Lee S, Chan L. Pneumococcal septicemia in children with sickle cell anemia. Changing trend of survival. JAMA. 1981; 245:1839–42.. [PubMed: 7230369]
3. Vichinsky E, Hurst D, Earles A, Kleman K, Lubin B. Newborn screening for sickle cell disease: effect on mortality. Pediatrics. 1988; 81:749–55. [PubMed: 3368274]
4. King L, Fraser R, Forbes M, Grindley M, Ali S, Reid M. Newborn sickle cell disease screening: the Jamaican experience (1995–2006). J Med Screen. 2007; 14:117–22. [PubMed: 17925083]
5. Yanni E, Grosse SD, Yang Q, Olney RS. Trends in pediatric sickle cell disease-related mortality in the U.S., 1983–2002. J Pediatr. 2009; 154:541–5. [PubMed: 19028391]
6. Lee A, Thomas P, Cupidore L, Serjeant B, Serjeant G. Improved survival in homozygous sickle cell disease: lessons from a cohort study. Br Med J. 1995; 311:1600–2. [PubMed: 8555802]
7. Serjeant GR, Serjeant BE. Management of sickle cell disease; lessons from the Jamaican Cohort Study. Blood Rev. 1993; 7:137–45. [PubMed: 8241829]
8. King L, Ali S, Knight-Madden J, MooSang M, Reid M. Compliance with intramuscular penicillin prophylaxis in children with sickle cell disease in Jamaica. West Indian Med J. 2011; 60:177–80. [PubMed: 21942123]
9. Hardie R, King L, Fraser R, Reid M. Prevalence of pneumococcal polysaccharide vaccine administration and incidence of invasive pneumococcal disease in children in Jamaica aged over 4

years with sickle cell disease diagnosed by newborn screening. Ann Trop Paediatr. 2009; 29:197–202. [PubMed: 19689861]