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 specific interventions such as penicillin prophylaxis or pneumococcal conjugate vaccines (PCVs). Three references were cited describing reductions in SCD mortality that occurred prior to the availability of those interventions. 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. 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. 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.

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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.\(^5,7\) 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

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