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INAUGURAL NEWBORN SCREENING FOLLOW-UP BRIEF

SICKLE CELL DISEASE IN MICHIGAN

BACKGROUND

Sickle cell disease is an inherited disorder that affects the formation of hemoglobin, the protein in red blood cells that carries oxygen.[1,2] The abnormal shape of red blood cells found in patients with sickle cell disease contributes to co-morbidities throughout the life span including pneumococcal infections and acute spleen sequestrations in infants[3] and renal medullary carcinoma in young adults.[4] Sickle cell disease is also associated with premature mortality.[5]

A 1986 study demonstrated that penicillin prophylaxis markedly reduces the incidence of pneumococcal sepsis among children (<three years of age) with sickle cell anemia.[6] This study provided powerful incentive for the widespread implementation of newborn screening (NBS) for sickle cell disease. Additionally, when linked to timely diagnostic testing, parental education, family counseling, and comprehensive care, NBS notably reduces morbidity and mortality associated with sickle cell disease in infancy and early childhood.[7] In 1987, the State of Michigan passed Public Act 14, which established statewide screening of all newborns for sickle cell disease. The Act also mandated a fee to fund the program and added comprehensive programs for follow-up, medical management, and quality assurance.

Following the cohort of sickle cell disease patients diagnosed through NBS in Michigan is challenging. However, we can utilize statewide databases to evaluate the prevalence, morbidity, and mortality of sickle cell disease across the life span.

STATEWIDE DATABASES

The NBS Follow-up Program maintains a database of initial screening results and data received from the Sickle Cell Disease Association of America (SCDAA)-Michigan Chapter on patients referred for follow-up. The SCDAA sends information about confirmatory results, treatment initiation, and dates of clinic visits. We used this database to examine the cumulative prevalence of sickle cell disease among newborns since screening began and to provide more detailed information on the 2008 birth cohort of cases.

The Michigan Inpatient Database (MIDB) is a database of hospital information collected by the Michigan Health & Hospital Association (MHA). Each hospital in the state reports data to the MHA, and the MHA aggregates the reported data. The Michigan Department of Community Health purchases the MIDB from the MHA. We used 2007 MIDB information to determine the number of hospitalizations for patients with sickle cell disease and the demographics of those patients. We also examined diagnostic and procedural codes for the subset of patients with a primary diagnosis of sickle cell disease to assess common co-morbidities and services utilized by hospitalized patients with sickle cell disease.

Death certificate data containing race, birth date, death date, and causes of death for all people who died in Michigan from 1970-2004 with sickle cell disease or trait listed as a cause of death was used to assess mortality. This data was restricted to deaths among blacks since 99.1% of the deaths occurred among black people. We examined the age distribution of the deaths and used generalized linear modeling (GLM) to determine the relation between year of death and age at death. We used census population data to estimate the sickle cell mortality rate (MR) and GLM to assess the change in MR over time.

WHAT DO WE KNOW ABOUT THOSE WITH SICKLE CELL DISEASE?

Since screening for sickle cell disease began in 1987, 1,436 confirmed cases have been identified in Michigan, resulting in a cumulative detection rate of 1:2,074 newborns screened. In 2008, 47 newborns were diagnosed with sickle cell disease; the detection rate was 1:2,539 newborns screened (Table 1). Of patients with sickling conditions in the 2008 birth cohort, 66% had sickle cell anemia, 23% had SC disease, and the remaining 11% had sickle β thalassemia. Black infants accounted for 87% of the cases in 2008. Thus, the detection rate in black infants was 1:542 newborns screened, nearly 5 times the overall detection rate.

In 2007, sickle cell disease was recorded as a diagnosis in 4,570 hospital stays in Michigan. Table 2 describes the hospitalization characteristics

Table 1. Sickle Cell Disease Screening, Michigan, 2008

Disorder	Confirmed + (N)		Detection Rate	
	Total	Among Blacks	Total*	Among Blacks*
Sickle cell anemia	31	27	1:3,849	1:795
SC disease	11	9	1:10,848	1:2,385
Sickle β thalassemia	5	5	1:23,865	1:4,293
Total	*47*	*41*	*1:2,539*	*1:524*

**Out of the number of Michigan resident infants screened, total N=119,327, among Blacks N=21,465*

where sickle cell disease was the primary diagnosis or sickle cell disease was listed as any diagnosis. Of all hospital stays with any sickle cell disease diagnosis, approximately 68% had a primary diagnosis of sickle cell disease. Over 85% of hospitalizations for sickle cell disease occurred among black patients. Of all stays, 70.1% occurred among adults (18 years+).

An estimated 79% of children and 93% of adults with a primary diagnosis of sickle cell disease had a secondary diagnosis recorded. The most common associated condition for children was fever (21%), while the most common for adults was pneumonia (5%). The majority of children (81%) and adults (58%) did not have any procedures recorded for their hospitalization. Transfusion of packed cells was the most common primary procedure for both children and adults, accounting for over half of recorded primary procedures.

From 1970-2004, 900 deaths occurred in Michigan where sickle cell was listed as a cause of death. The average age at death was 34 years old and the range was 1 day of life to 96 years. Average age at death significantly increased from 1970-2004 ($p<0.001$) (Figure 1). The mean sickle cell mortality rate (MR) from 1970-2004 was 20.4 deaths/1 million black residents, and the MR did not significantly change over time.

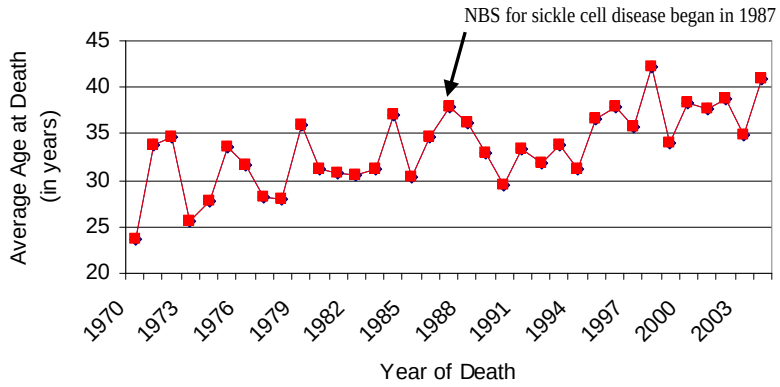

Figure 1. Average Age at Death by Year of Death for People with Sickle Cell, Michigan, 1970-2004

Table 2. Demographics of Hospitalizations of Patients with Sickle Cell Disease, Michigan, 2007

		Principal diagnosis of sickle cell disease	Any diagnosis of sickle cell disease
All discharges		3134 (100%)	4570 (100%)
Age groups	<1 year	32 (1.0%)	89 (2.0%)
	1-17 years	938 (29.9%)	1276 (27.9%)
	18-44 years	1852 (59.1%)	2572 (56.3%)
	45-64 years	302 (9.7%)	559 (12.2%)
	65 years +	10 (0.3%)	74 (1.6%)
Sex	Female	1516 (48.4%)	2410 (52.7%)
	Male	1618 (51.6%)	2160 (47.3%)
Race	Black	2790 (89.0%)	3948 (86.4%)
	White	8 (0.3%)	70 (1.5%)
	Other	30 (1.0%)	49 (1.1%)
	Missing	306 (9.7%)	503 (11.0%)

CONCLUSIONS AND FUTURE DIRECTIONS

The Michigan NBS Follow-up Program utilized NBS data, data from the SCDAA, and other databases to assess the prevalence of sickle cell disease, co-morbidities, and mortality across the life span. The average age at death for people with sickle cell disease has increased since NBS for sickle cell disease began in 1987. However, more research is needed to tease out the effects of early identification and intervention due to NBS from those of advancing medical practices.

The Follow-up Program plans to collaborate with other state programs to continue assessing the prevalence of sickle cell disease for all ages and health outcomes of people with sickle cell disease in Michigan.

ABOUT NEWBORN SCREENING FOLLOW-UP IN MICHIGAN

The Michigan Newborn Screening Program screens newborns in the state for 49 disorders, including sickle cell disease. The screening is performed within the Division of Chemistry and Toxicology in the Bureau of Laboratories. The NBS Follow-up Program, located in the Division of Genomics, Perinatal Health and Chronic Disease Epidemiology within the Bureau of Epidemiology, oversees short and long-term follow-up of infants identified through the program. Follow-up begins with referring these infants to one of four NBS-funded medical management centers for diagnosis and treatment. The Follow-up Program maintains short and long-term follow-up databases for monitoring and evaluation. Education, training, and quality assurance measures are also responsibilities of the Follow-up Program.

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