

## **ORAL PENICILLIN PROPHYLAXIS IN CHILDREN WITH SICKLE CELL ANEMIA**

Children born with sickle cell anemia have increased susceptibility to severe bacterial infection that can lead to illnesses such as meningitis, pneumonia, or septicemia. Such infections are a major cause of death among these children, especially those under three years of age, and have been documented to occur in children as young as four months of age. *Streptococcus pneumoniae*, particularly, is a significant pathogen for individuals with sickle cell anemia. The nationwide Cooperative Study of Sickle Cell Disease found the annual incidence of pneumococcal septicemia in sickle cell anemia patients to be 10 per 100 persons years with a case fatality rate of 30% (Gaston et al., New England Journal of Medicine, Vol. 134, p. 1593, June 19, 1986.)

In 1983, the Sickle Cell Disease Branch of the Division of Blood Diseases and Resources, National Heart, Lung, and Blood Institute, established the Prophylactic Penicillin Study to assess the efficacy of oral penicillin in preventing severe bacterial infections (bacteremia, meningitis, and pneumonia requiring hospitalization) in children with sickle cell anemia. The study effectively demonstrated the value of oral penicillin in preventing pneumococcal septicemia in children with sickle cell anemia. The results were so pronounced in favor of the penicillin treatment over the placebo that the trial was terminated eight months before its planned ending date.

Based upon the findings and implications of the Prophylactic Penicillin Study, the Sickle Cell Disease Branch of the national Heart, Lung, and Blood Institute is disseminating the following recommendations:

1. Infants of high risk populations should be screened for sickle cell anemia in the newborn period.
2. Those diagnosed with **sickle cell anemia** should be placed on **oral prophylactic penicillin** 125 mgs B.I.D. by the age of **four months**.

Oral prophylactic penicillin should be used in conjunction with the standard immunization and treatment protocol set out in the National Institutes of Health Publication number 85-2117, Management and Therapy of Sickle Cell Disease.

In the State of New York, all newborns are screened for Sickle Cell Anemia through the Department of Health's Newborn Screening Program (Public Health Law 2500-a and 10 NYCRR Part 69). Children who were not born in New York State and/or whose sickle cell status is unknown should be screened as soon after birth as possible.

The age at which prophylactic penicillin can safely be discontinued remains to be clearly defined. Premature cessation of prophylactic penicillin therapy might expose children to increased risk of *streptococcus pneumoniae* infection if cessation occurs prior to development of adequate immunity. For this reason, experts recommend that **penicillin prophylaxis be continued for five years, at a minimum**. (A collaborative study will soon be undertaken by the Sickle Cell Branch of the National Heart, Lung, and Blood Institute in order to determine the safest age at which to discontinue penicillin prophylaxis.) It is important to remember that the dose should be adjusted as the child ages and/or gains weight. Children with penicillin allergy should be placed upon an equivalent dose of the most appropriate antibiotic which has a spectrum similar to that of penicillin. (The efficacy of antibiotics other than penicillin were not evaluated in the penicillin prophylaxis study.)