

# Sickle Selections

a quarterly newsletter from the University of Rochester Sickle Cell Program

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## Sickle Cell Disease: What to Expect in the First Ten Years

The Cooperative Study of Sickle Cell Disease has reported\* on 694 infants with confirmed sickle cell disease followed for their first decade. In all forms of sickle cell disease, painful crises and acute chest syndrome were the most common sickle-related events. The half-time to the first painful crisis was 4.9 years in SS disease but 7.1 years in SC disease. The acute chest syndrome was experienced by half of all SS patients by 5.8 years. Hand-foot syndrome occurred in 25% of SS patients by 2.5 years, but was uncommon in SC patients. Bacteremia occurred most commonly under 4 years of age in SS children, but under 2 years of age in SC children.

Of the 694 children, 20 died during the 10-year study period, a mortality rate of 2.9%. All of the deaths were in the SS group. Eleven died of infection (9 *S. pneumoniae* and 2 *H. influenzae*), 1 from a CVA, 2 from splenic sequestration, and 9 were undiagnosed. It was not clear to what extent noncompliance with penicillin prophylaxis was responsible for the infectious deaths.

\* Gill FM et al. *Blood* 86:776-783, 1995.

## Delayed Adolescent Growth in Homozygous Sickle Cell Disease

A study reported by A.Singhal in the *Arch Dis Child*\*\* reports longitudinal observations of height in a Jamaican cohort of children with homozygous sickle cell disease (SS), sickle cell hemoglobin C (SC) dis-

ease, and normal (AA) hemoglobin. Subjects were identified by neonatal screening of 100,000 consecutive deliveries at a major maternity hospital in Jamaica. Analysis included 44 SS patients, 44 SC patients and 44 AA control children. Height was measured at 3 month intervals for SS and SC patients and every 6 months for the AA control children.

The data demonstrate that SS patients have a 1.4 year delay in mean age of initiation of the adolescent growth spurt, a 1.6 year delay in mean age at peak height velocity, and a lower height velocity at the time of the onset of the growth spurt compared with AA controls.

The first pubertal changes occurred at  $12.8 \pm 1.6$  years in SS males and  $12.0 \pm 1.8$  years in SS females, compared with  $11.1 \pm 1.2$  years in AA males and  $10.1 \pm 1.2$  years in AA females.

The age of menarche in SS girls was significantly later than in AA girls ( $15.4 \pm 1.3$  years vs  $13.1 \pm 1.3$  years;  $P < 0.001$ ).

	Delay in		Age of Menses
	growth spurt	age at peak ht velocity	
SS	1.4 yrs	1.6 yrs	15.4
SC	no delay	—	13.7
AA			13.1

\*\* Singhal A. et al. *Arch Dis Child* 1994; 71:404-408

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