

Sickle Selections

a quarterly newsletter from the University of Rochester Sickle Cell Program

April 1997

Acute Chest Syndrome (ACS) in Sickle Cell Disease

A recent article in Blood¹ by The Cooperative Study of Sickle Cell Disease has prospectively followed 3,751 patients from birth to 66 years of age for ACS. There were 1,722 ACS episodes in 939 patients.

Young children (age 2 to 4 years) presented with fever and cough, a negative physical exam, and rarely had pain. Adults were afebrile and complained of shortness of

ACS is the second most common cause of hospitalization of sickle cell disease patients and is responsible for up to 25% of deaths.

breath, chills, and severe pain. *Upper lobe disease was more common in children; multilobe and lower lobe disease affected adults more often. Bacteremia was documented in 3.5% of episodes, but was strongly influenced by age (14% of infants and 1.8% of patients > 10 years). ACS was most common in winter with children having the most striking increase. Fifty percent of adults had a pain event in the 2 weeks preceding ACS and children were more likely to have febrile events.*

The death rate was four times higher in adults than in children. Fatal cases generally developed rapid pulmonary failure and one third were associated with bacteremia. Age has a striking effect on the clinical picture of ACS. *In children, ACS was milder and more likely due to infection, whereas in adults ACS was severe, associated with pain and had a higher mortality rate.*

¹ Blood, Vol 89, No5 (March 1), 1997:pp 1787-1792

Birth Statistics² for Monroe and Surrounding Counties³ in NY

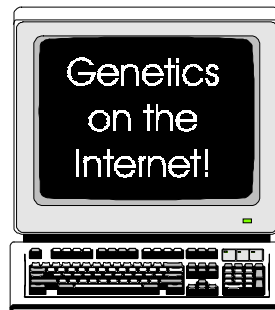
	1994	1995	1996
Race/ethnicity⁴			
White	13,730	13,297	
Afr-Amer	2359	2106	
Hispanic	746	721	
Other	466	472	
Total	17,301	16,596	
Disease⁵			
S/S	5	4	1
S/C	4	6	2
S/Thal	1		
S/OArab	1		
Total	11	10	3
Trait			
A/S	217	215	191
A/C	64	61	58
A/OArab	2	1	1
Total	283	277	250

² NY State Department of Health

³ Chemung, Livingston, Ontario, Schuyler, Seneca, Stueben, Wayne, Yates

⁴ No statistics available for 1996

⁵ All disease babies born to African American women



We are considering creating a site on the internet to provide information on genetics. We would appreciate your answering the questions on the stamped postcard enclosed and mailing it back to us.

Thank You!

UNIVERSITY OF
ROCHESTER
MEDICAL CENTER
Strong Sickle Program
Offers Help

Regarding treatment contact:

Dr. Norma Lerner or Pat Lamarche R.N., P.N.P. Department of Pediatrics 275-2981
Dr. Karen Kaplan, Department of Medicine 275-3761

Regarding laboratory diagnosis, newborn screening and genetic counseling, contact:

Dr. Peter Rowley, Sandra LaBella or Starlene Loader, Division of Genetics 275-4602

Visit our website: www.urmc.rochester.edu/genetics