

# 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 *Blood*<sup>1</sup> 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.*

<sup>1</sup> *Blood*, Vol 89, No5 (March 1), 1997:pp 1787-1792

## Birth Statistics<sup>2</sup> for Monroe and Surrounding Counties<sup>3</sup> in NY

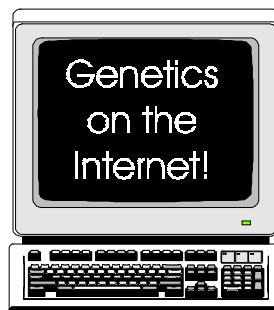
	1994	1995	1996
<b>Race/ethnicity<sup>4</sup></b>			
White	13,730	13,297	
Afr-Amer	2359	2106	
Hispanic	746	721	
Other	466	472	
<b>Total</b>	<b>17,301</b>	<b>16,596</b>	
<b>Disease<sup>5</sup></b>			
S/S	5	4	1
S/C	4	6	2
S/Thal	1		
S/OArab	1		
<b>Total</b>	<b>11</b>	<b>10</b>	<b>3</b>
<b>Trait</b>			
A/S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

<sup>2</sup> NY State Department of Health

<sup>3</sup> Chemung, Livingston, Ontario, Schuyler, Seneca, Stueben, Wayne, Yates

<sup>4</sup> No statistics available for 1996

<sup>5</sup> 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

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