

Sickle Selections

a quarterly newsletter from the University of Rochester Sickle Cell Program

April 1999

Newborn with SE Disease Identified

NY State Newborn Screening has identified a newborn with hemoglobin S/E disease. This is the first such reported case in our 9 county area since we began receiving reports of sickle cell disease births in 1975. The African-American father transmitted the sickle gene and the Cambodian mother transmitted the hemoglobin E gene. The few cases for hemoglobin S/E disease reported in the literature describe a mild anemia with slight splenomegaly. This newborn has been placed on oral prophylactic penicillin and the mother has been instructed in the signs and symptoms of sickle cell disease and the need to seek prompt medical attention if signs of infection or anemia develop.

References:

1. Lachant, N.A. *Amer. Journal of Hematology* 1987; 25: p449
2. Vishwanathan C. *Indian Pediatrics* July 1992; 29: p 895

Summer Camps for Children with Sickle Cell Disease

Please share this information with your sickle cell families.

CAMP OPEN ARMS, sponsored by Cancer Action, Inc. is offering a summer day camp for children, ages 3 to 14, with cancer or other blood diseases, including sickle cell. Siblings are also invited. The camp runs from **July 6-16, 1999**, and will be located in Penfield at Cobbles Elementary School. **Transportation will be provided.** Applications may be obtained by calling Cancer Action at (716) 423-9700. They are due May 31, 1999

CAMP GOOD DAYS AND SPECIAL TIMES, along with the Rochester Sickle Cell Support Group, is planning a Family Day on Saturday, Aug. 7, 1999 at the camp on Keuka Lake. The goal of this day is to give families of children with sickle cell disease a chance to have fun together and to survey the community about establishing a program for children with sickle cell disease at Camp Good Days. **Transportation will be provided.** For further information, please contact Edwina Daniels (787-0838) or Eva Stanley (482-4941).

Birth Statistics¹ for Newborns in HSA Region II of NY State²

The New York State Newborn Screening Program requires a blood specimen from every baby born in the state. Among the 7 disorders tested for by the state on these specimens are hemoglobinopathies. A list of the hemoglobinopathies identified in newborns in HSA region II over the past 3 years is listed below.

	Disease			Trait			
	1998	1997	1996	1998	1997	1996	
S/S	5	2	1	A/S	254	233	192
S/C	1	4	2	A/Fast	57	60	56
S no A ³	2	4		A/C	53	57	57
S/β ⁺ -Thal	1			A/E or O	11	19	9
S/E	1			A/D or G	6	5	13
E/E	1	1		A/Fetal	4	4	7
C no A ³			1	A/O Arab	1	3	1
Total	11	11	4	A/Variant			1
				Total	386	381	336

Incidence in Black Births

	NY State ⁴	Monroe County ⁵
Sickle Disease (S/S, S/C, S/OArab, S/Thal)	1/240	1/196
Trait (A/S, A/C, A/OArab)	1/9	1/8

¹ Source: NY State Department of Health

² Chemung, Livingston, Monroe, Ontario, Schuyler, Seneca, Stueben, Wayne, Yates

³ An abnormal hemoglobin and no hemoglobin A can indicate either a homozygote for the abnormal hemoglobin or a compound heterozygote for the abnormal hemoglobin and β⁰-thalassemia. The best way to distinguish these is to test both parents.

⁴ For 1998

⁵ Average of years 1990-1996

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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