

Sickle Selections

a quarterly newsletter from the University of Rochester Sickle Cell Program

October 1997

Stroke Prevention in Sickle Cell Patients

In September, the National Heart, Lung, and Blood Institute (NHLBI) announced the important results of its **Stroke Prevention Trial in Sickle Cell Anemia (STOP)**¹. This study involved 130 sickle hemoglobinopathy patients who were considered to be at high risk for stroke on the basis of elevated cerebral blood flow velocity as measured by transcranial Doppler. The subjects, ages 2 to 16 years were from 14 different clinical trial centers and were randomized to either chronic blood transfusion therapy to maintain hemoglobin S under 30%, or to supportive care. After one year, ten children in the standard care group and one child in the transfusion group experienced a cerebral infarction. This result represented a 90% decrease in the stroke rate in the transfused group. In view of

A 90% decrease in the stroke rate in the transfused group over the standard care group was realized.

these compelling results, the study was terminated early so that children receiving supportive care could be offered transfusion therapy to prevent stroke.

The Pediatric Hematology Division at The Children's Hospital at Strong is presently in the process of establishing a transcranial Doppler screening program comparable to that used in the study. Our plan is to screen children with sickle syndromes from age 2 years onward and to make transfusion therapy available to those with studies showing a cerebral blood velocity greater than 200 cm/sec on two separate occasions. Rescreening will be obtained every 6 months until an optimal schedule of transfusion is found by further collaborative studies.

Our hope is to have a program in place by early spring of 1998. Please feel free to contact the doctor or nurse at the Comprehensive Sickle Cell and Hemoglobinopathy Center at Strong Memorial Hospital should you wish to study your patient or to obtain further information.

¹ Clinical Alert: (NHBLI) September 18, 1997

Flu Shots Recommended for Sickle Cell Children

As the winter months approach, so do concerns regarding influenza. The Pediatric Hematology Clinic is recommending that children greater than 6 months of age, with either sickle cell anemia or thalassemia, be immunized in preparation for this winter's influenza season.

According to the Red Book, influenza vaccine may be administered simultaneously with MMR, (but at a separate site and with a different syringe), Hemophilus b, pneumococcal and oral poliovirus vaccines. Since both influenza and pertussis vaccines in young children can cause febrile reactions, the Red Book recommends that influenza vaccine should not be given within 3 days of vaccination with DTP.

Annual vaccination is recommended because of declining immunity in the year after vaccination.

Incidence of Sickle Cell Disease by Racial/Ethnic group²

African-Americans	1/375	SS
	1/835	SC
	1/1667	S/βthal
Hispanics (Eastern states)	1/1100	SS, SC & S/βthal
Native Americans	1/2700	SS, SC & S/βthal
Asians	1/11,500	SS, SC & S/βthal
Hispanics (Western states)	1/32,000	SS, SC & S/βthal
Whites	1/58,000	SS, SC & S/βthal

²Pediatrics Vol 98, No 3, Sept 1996, p 491

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