

Sickle Selections

a quarterly newsletter from the University of Rochester Sickle Cell Program

January 1998

Cerebrovascular Accidents (CVA) in Sickle Cell Disease

The rates and risk factors of CVA in patients with sickle cell disease have been recently reported by The Cooperative Study of Sickle Cell Disease¹. Clinical data was collected on 4,082 sickle cell disease patients enrolled from 1978 to 1988. This study utilized a larger patient sample than previous studies.

Rates

The highest CVA prevalence (4.01%) and incidence (0.61 per 100 patient years) were in sickle cell anemia (SS) patients but CVA occurred in all common sickle genotypes.

Children less than 2 years of age had the lowest CVA incidence. However, the incidence was higher in the 1 to 9 years age group than in the 10 to 19 year age group.

Number of CVAs /100 patient-years

Age (yr.)	SS	SC	S-β ⁺	S-β ⁰	Total
N	2436	839	188	184	3647
<2	0.13(1)	0.00	0.00	0.00	0.08 (1)
2-5	1.02 (20)	0.27 (2)	0.00	0.00	0.75 (22)
6-9	0.79 (15)	0.00	0.00	0.00	0.55 (15)
10-19	0.41(15)	0.09 (1)	0.00	0.00	0.30 (16)
20-29	0.52 (14)	0.16 (1)	0.46(1)	0.43 (1)	0.45 (17)
30-39	0.59 (8)	0.00	0.00	0.00	0.39 (8)
40-49	0.74 (3)	1.01 (2)	0.00	0.00	0.76 (5)
>50	1.28 (2)	0.76 (1)	0.00	0.00	0.91 (3)
Overall	0.61 (78)	0.17 (7)	0.11 (1)	0.10 (1)	0.46 (87)
Age-adjusted	0.61	0.15	0.09	0.08	

The number in parentheses represents the number of events.

In SS patients the incidence of **infarctive CVA** was lowest in patients 20-29 years of age and **higher in children** and older patients. In contrast, the incidence of hemorrhagic stroke was highest among patients aged 20-29 years. Across all ages the mortality rate was 26% in the 2 weeks after hemorrhagic stroke. No deaths occurred after infarctive stroke. (Infarctive and hemorrhagic strokes were distinguished by imaging studies.)

1. Ohene-Frempong, Kwaku et al Blood, Vol 91, No 1 1998: pp288-294

2. Clinical Alert: (NHBLI) September 18, 1997

Risks

Risk factors for **infarctive stroke** include:

- prior transient ischemic attack (TIA)
- low steady-state hemoglobin concentration
- rate of and recent episode of acute chest syndrome
- elevated systolic blood pressure.

Risk factors for **hemorrhagic stroke** include:

- low steady-state hemoglobin concentration
- high leukocyte count

The chances of having a **first CVA** by:

	SS	SC
20 yrs	11%	2%
30 yrs	15%	4%
45 yrs	24%	10%

Prevention

For the last 2 decades, management of CVA in sickle cell disease has focused on prevention of recurrence but now there is a strong interest in preventing the first CVA. Predicting CVA is therefore important. Transcranial Doppler ultrasonography has been shown to select patients at high risk of stroke.² Chronic transfusion therapy can be offered to those patients.

The Pediatric Hematology Division at The Children's Hospital at Strong is presently establishing a transcranial Doppler screening program. We plan to have a program in place by the spring of 1998. You may contact Dr. Norma Lerner at the Comprehensive Sickle Cell and Hemoglobinopathy Center at Strong Memorial Hospital to obtain further information.

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Offers Help

Regarding treatment contact:

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

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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