

Sickle Selections

a quarterly newsletter from the University of Rochester Sickle Cell Program

July 2001

Highlights from the National Sickle Cell Disease Program Conference of April 2001

We report here on Transcranial Doppler Ultrasonography. We will report on Hydroxyurea in the next newsletter.

Transcranial Doppler Ultrasonography (TCD)

Investigators at the Children's Hospital of Philadelphia reported on a 2 year follow-up study of a cohort of children with sickle cell disease. These children, like those cared for at the Children's Hospital at Strong, had received TCD screening to assess cerebral artery blood flow velocity. (Screening is now considered standard of care for children with sickle cell disease since studies have shown that abnormal velocities are associated with a significantly increased risk of stroke.^{1,2}) A normal exam is defined as all velocities < 170 cm/sec; conditional exam 170 to 200 cm/sec; an abnormal exam > 200cm/sec in either the internal carotid or the middle cerebral artery.

TCD exams were performed on 235 children with a mean age of 9.4 y (range 2-18.9 y). The initial TCD was normal in 81%, conditional in 14%, and abnormal in 5% of the children. 74 of the 191 children with normal studies had follow-up studies. 88% remained normal, 8% became conditional, and 4% became abnormal. 25 of 32 children with conditional studies had follow-up. 32% became normal, 36% remained conditional, and 32% became abnormal. For children with initially conditional studies, the mean velocity was higher for those that went on to develop abnormal studies then for those that remained the same or improved. Children whose TCD study classification worsened were younger than those whose studies remained the same or improved (mean 6.8 y vs. 9.2 y). Since none of the children with initially normal or conditional studies developed a clinical stroke, follow-up intervals were felt to be appropriate. (Notably those children with abnormal studies were placed on prophylactic chronic transfusion therapy.)

The study concluded that more frequent TCD screening might be advisable for children with conditional studies, particularly those in the in the high conditional range, because of the high rate of change to abnormal studies. More frequent screening of younger children was also felt to be a future consideration.

1. N Engl J. Med 1992; 326: 601-610.

2. N Engl J. Med 1998; 339: 5211.



The Children's Hospital at Strong currently recommends:

- All children with sickle cell disease over the age of 2 years should receive TCD screening on an annual basis. Conditional studies should be repeated every 3 months and abnormal studies within 6 weeks.
- Chronic prophylactic transfusion should be offered to those patients with 2 successive abnormal studies, as is recommended by the National Institutes of Health.

To arrange a study for your patient call Patricia Lamarche PNP at 275-2981.



Summer Camp for Children with Sickle Cell Disease

Camp Kope the special summer camp for children with sickle cell disease will be held July 6-8 at Camp Good Days and Special Times on Keuka Lake.

If you have families that would benefit from this summer program have them call the Camp Good Days office at 624-5555 for a brochure and registration form.



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