

Optimizing Care in Sickle Cell Patients on Chronic Exchange Transfusion Therapy

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Disclosures

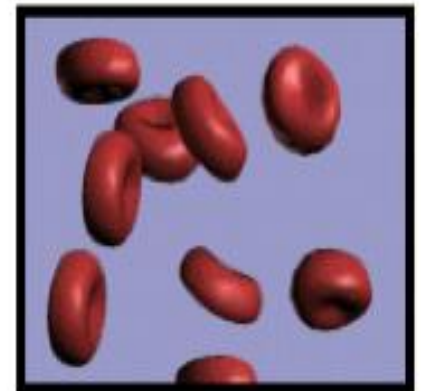
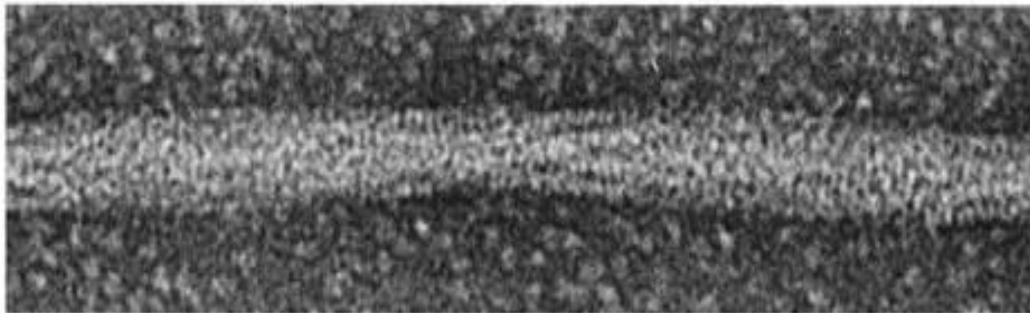
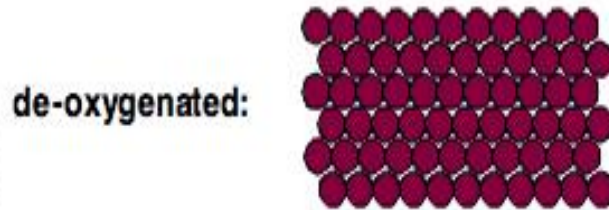
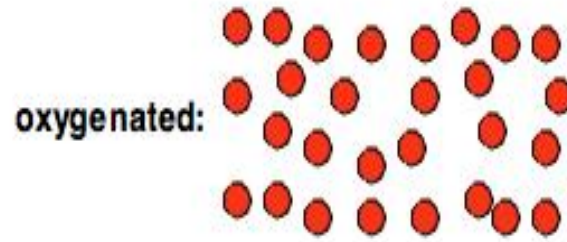
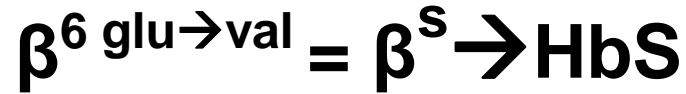
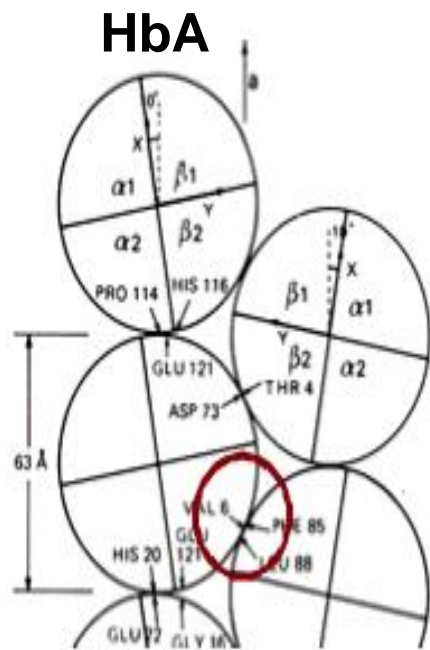
We have no relevant financial relationships with commercial interests:

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- **Phandee Watanaboonyongcharoen MD**

Agenda

- **Describe sickle cell disease, etiopathogenesis and complications**
- **Rationale for chronic exchange transfusion therapy in sickle cell disease**
- **Description of the study**

What is Sickle Cell Disease?

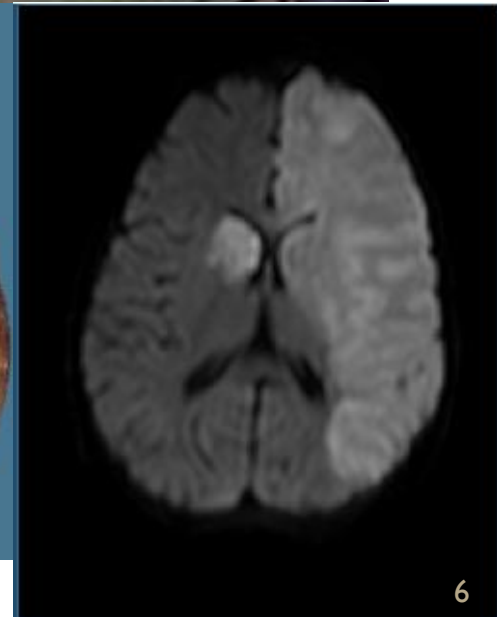


Demographics

- **Common in people from Sub Saharan Africa, Central America, Caribbean, Mediterranean and Indian descent**
- **Sickle cell disease:50,000-70,000 population affected**
 - **African Americans: 1:500 births**
- **Sickle cell trait: 1:12 African-Americans**

Sickle cell symptoms

- **Chronic hemolytic anemia**
- **Acute Complications**
 - Acute pain, dactylitis
 - Acute stroke
 - Cholecystitis
 - Hepatic/splenic sequestration
- **Chronic multiorgan damage**
 - Sickle cell retinopathy
 - Stroke
 - Autosplenectomy



**What is the Rationale
for using
transfusions in sickle
cell disease?**

Types of transfusions

Acute

- For acute stroke, acute chest, syndrome, multiorgan failure, hepatic/splenic sequestration

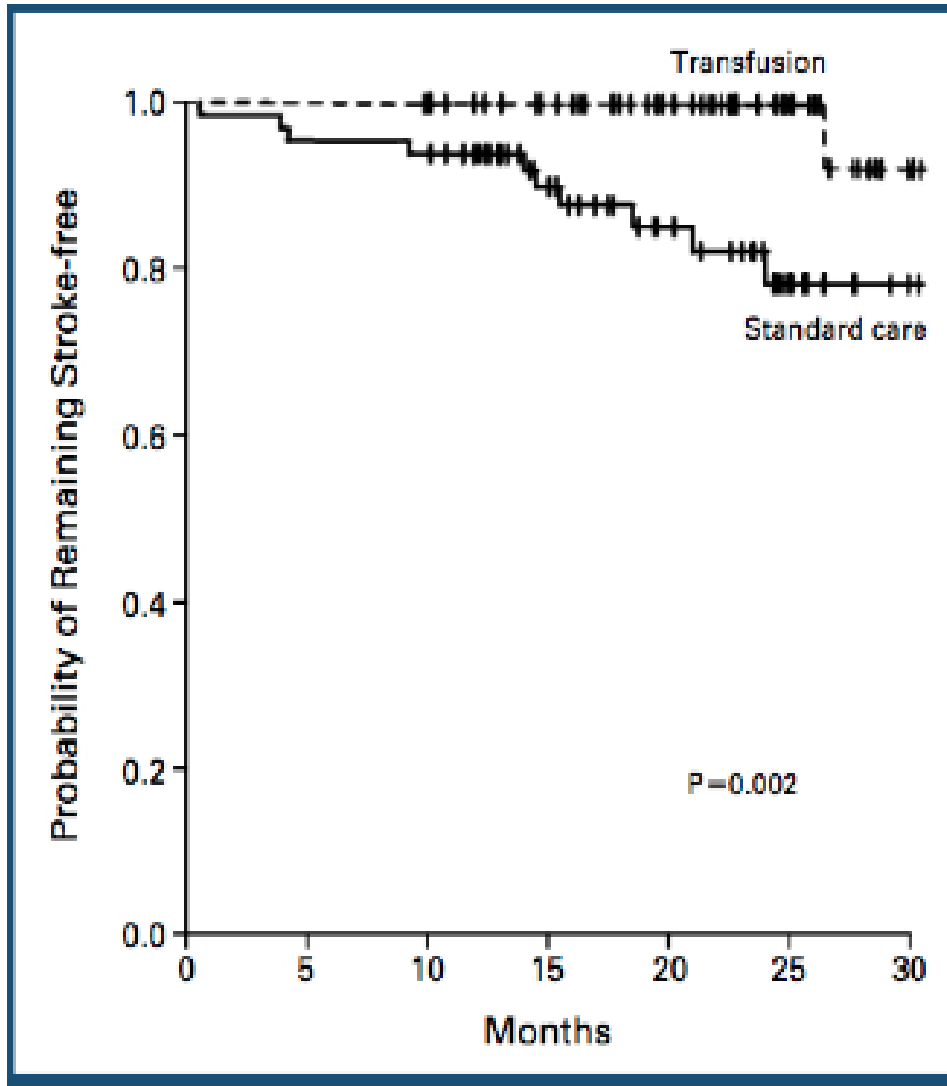
Chronic

- Exchange transfusions
 - For primary & secondary stroke prevention

Complications of Transfusions

- Iron overload
- Volume overload
- Alloimmunization
- Hyperviscosity
- Transfusion reactions
- Infections

STOP 1 TRIAL (1998)



- Sickle cell pts randomized to chronic transfusion vs standard care
- Chronic transfusions associated with >90% reduction of stroke risk
- Observed stroke rate = 10% per year over 2 years

Advantages of Chronic Exchange Transfusions

- **Performed at monthly intervals**
- **Reduced incidence of complications**

Objective

- **To optimize care of sickle cell patients on chronic exchange therapy at UNC**

METHODS

- **Retrospective analysis of SCD patients on chronic exchange therapy**
- **Number of patients: 24**
- **Timeline: data reviewed from 2009 - to date**
- **Reviewed:**
 - ❖ **Demographics**
 - ❖ **Indication for chronic exchange transfusion**
 - ❖ **Therapeutic orders**
 - ❖ **Volume of pRBCs needed**
 - ❖ **Adequacy of access**
 - ❖ **Adverse events**
 - ❖ **Compliance**
 - ❖ **Screening for iron overload, vitamin D deficiency & sickle cell retinopathy**

RESULTS (1)

- **SCD patients reviewed: n=24**
- **Age: 11 years to 36 years**
- **Gender: 16 males; 8 females**
- **Subtypes:**
 - ◆ **HbSS genotype = 22**
 - ◆ **HbSC genotype = 1**
 - ◆ **HbS β^0 genotype = 1**
- **Indication:**
 - ◆ **Post cerebrovascular event = 17**
 - ◆ **Transient ischemic attacks (TIAs) = 2**
 - ◆ **Recurrent Priapism = 2**
 - ◆ **Chronic & refractory pain = 3**

Results (2)

Category	Number of pts /total pts	Percentage of pts	Intervention
★★ Met Goals	10/24	42%	None
Hemodilution	2/24	8%	Replace with 5% albumin + NS
Compliance	21/24 > 85% 3/24 < 70%	87.5% 12.5%	Counselling
Access adequacy	23/24	96% with no access issues	1 pt needed line placement but lost to f/u
Adverse Events	4/24: allergic 7/24: citrate 2/24: febrile 6/24: Alloantibodies	17% = allergic 29%= citrate 8.3%= febrile	Supportive care & Provided antigen negative pRBC units



- 10 patients did not meet pre-ex HbA goal
- 1 patient did not meet any goals
- 3 patients did not meet post-ex Hct

Comprehensive Screening

Category	Number of pts /total pts	Percentage of pts	Intervention
★ Iron overload ferritin>1000ng/ml	5/24	21%	Chelation Rx Decrease post-ex HCT
Cerebral Imaging	18/19	95%	Annual MRI Brain
Viral Serologies (HBV, HCV, HIV)	Both: 9 HIV:1 HBV/HCV:4	38% 4% 17%	Annual serologies for frequent blood exposure
Vitamin D Levels (5-23 ng/ml)	Mild: 6 Mod:9 Sev:4	25% 42% 17%	Started Vitamin D supplements
Ophthalmology f/u	11/21	52%	For Sickle cell retinopathy

★ 2 patients s/p resolved iron overload > chelation

Conclusions

- Achievement of post exchange therapy goals
- Better documentation of medical records
- Efficient communication amongst patients, families and physicians
- Routine comprehensive screening
 - ❖ Quarterly ferritin levels
 - ❖ Ferritin >1000 ng/ml → chelation therapy
 - ❖ Annual MRI brain
 - ❖ Annual Ophthalmology follow up
 - ❖ Vitamin D levels and starting supplementation if deficient

ULTIMATE GOAL: For patients to obtain maximum benefit from this program!!!

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Acknowledgements

- **Araba Afenyi-Annan MD MPH**
- **Yara A Park MD**
- **Phandee Watanaboonyongcharoen MD**
- **UNC Apheresis team**
- **UNC Transfusion services**
- **Sickle cell patients**