Comprehensive Care of the Adult With Sickle Cell

Ward Hagar
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Director, Adult Sickle Cell Program
UCSF Benioff Children's Hospital Oakland

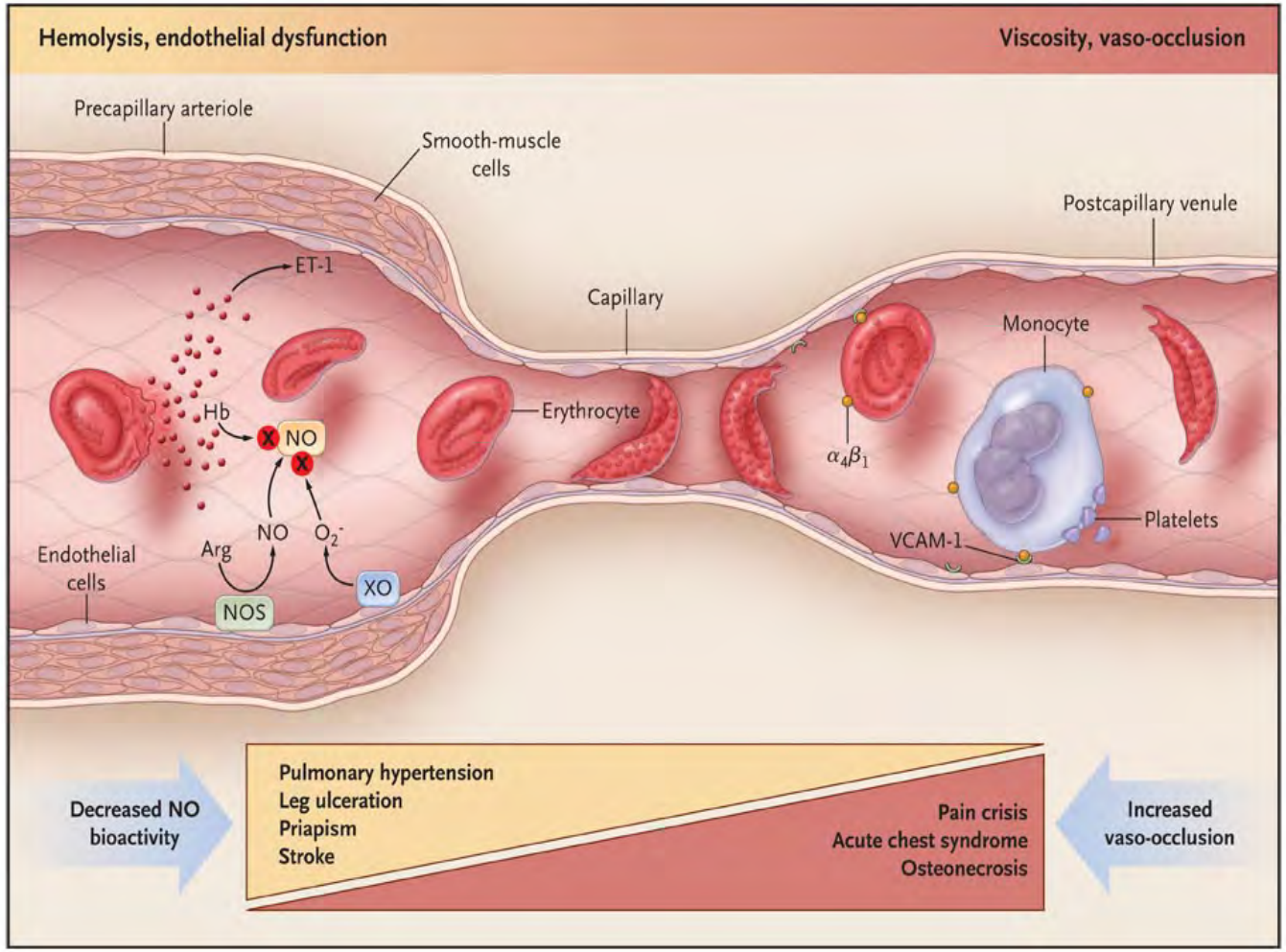
Talk Outline

- How Is Adult Care Different From Pediatric Care?
- What Are The Likely Problems That May Develop In Adults?
- Should We Be Screening For Possible Adult Problems?
- What Preventative Care Can Be Done For These Problems?

Big Picture

- Roughly 100,000 People With Sickle Cell In The USA
- More Adults Are Living With Sickling Diseases
- Sickle Cell Has Become A Chronic, Cumulative Disease
- Adults Are Also Developing “Usual” Adult Diseases
- Medical And Social Needs Are Increasing As Children Become Parents

Clinical Phenotypes of SCD



Gladwin M and Vichinsky E N Engl J Med 2008;359:2254

Mortality in Sickle Cell Disease from the Cooperative Study of Sickle Cell Disease

Methods: 3764 patients were enrolled from birth to 66 years of age in a prospective analysis

Results	Median age at death (yrs)	
	men	women
Sickle cell anemia (Hb SS)	42	48
Hb SC	60	68

Platt et al. N Eng J Med 1994;330:1639

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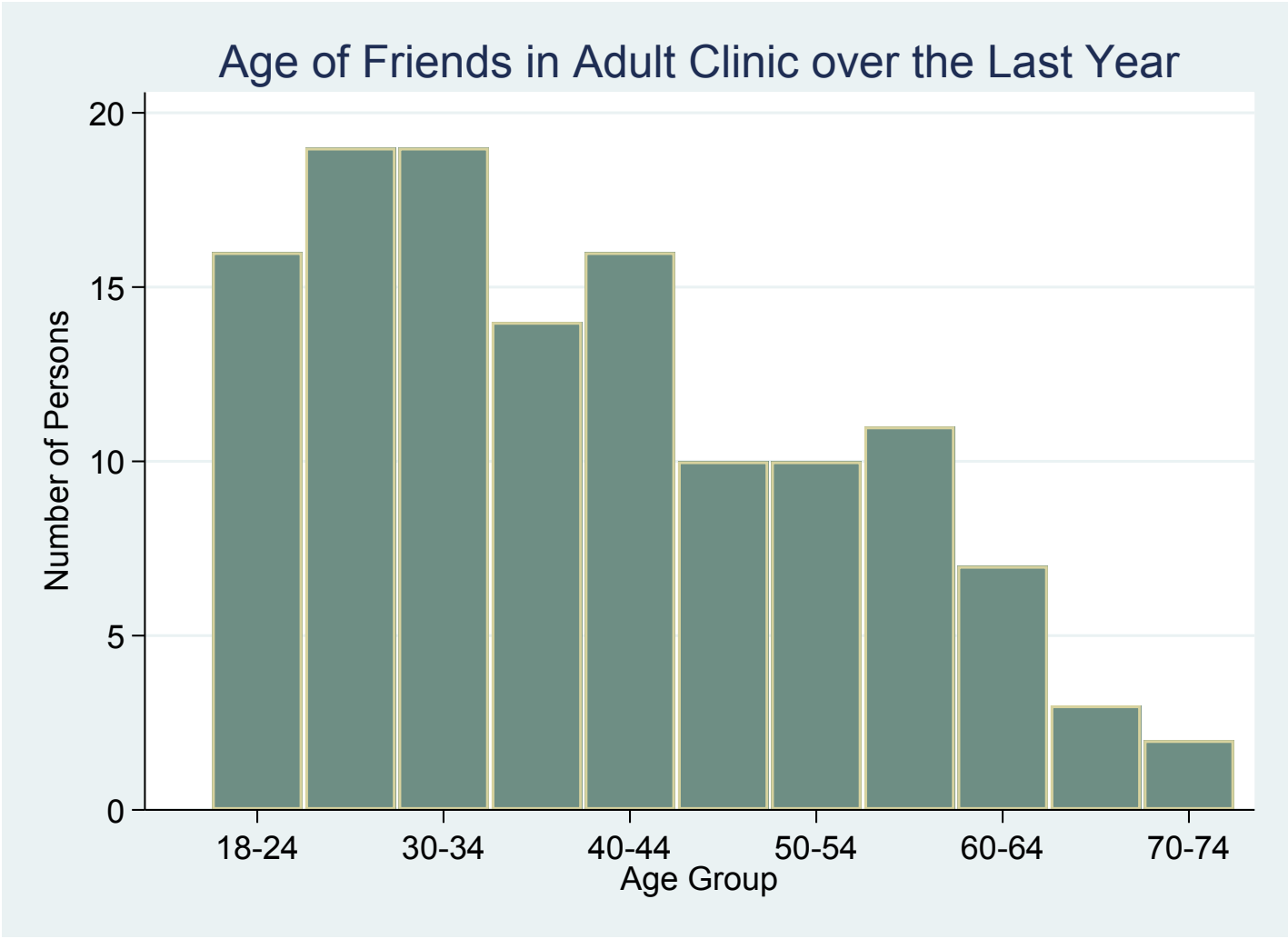


Challenges for the Northern Californian Patient

- Transportation
  - Patients moving farther from city centers
  - Price of transportation going up
- Referrals
  - Non-overlapping networks
  - Insurance issues
- Co-pays
  - Often prohibit treatments, consults, or medicine
- Follow-Up
  - Subtle and not so subtle social limitations
- Emergency Room Services
  - Often stigmatization and prolonged waits

How Is Adult Care Different From Pediatric Care?

- Many of the usual sickle cell serious events have not happened and are now less likely to happen
- Pain patterns have often been established
- Care is for more chronic issues and less for acute issues
- Adult diseases and sickle cell issues complicate each other
- Each adult bears more responsibility for their care and their health outcomes

What Are The Likely Problems That May Develop In Adults

1. Sickle cell and non-sickle cell conditions begin to run together

2. Chronic end organ dysfunction becomes the major factor in quality and quantity of life

3. General health care, as different from sickle cell specialty care, often becomes the determinant of longevity

4. Pain may become more chronic and less acute

Adult Issues Predominate

- Hypertension
  - Often with a family history
  - Any renal issues may worsen blood pressure
- Heart Disease
  - Becoming more frequent as everyone is “older”
  - Not clear if different from typical heart disease
- Renal Issues
  - Decreased function common, often not a clinic issue
  - Small subset go on to have more severe renal issues
- Liver Issues
  - Often iron overload related
- Bone Health
  - Avascular necrosis, vitamin D, and calcium issues.
- Eye Issues
  - Changes to small vessels of the eye
- Diabetes
  - Similar to background population
- Cancer screening
  - No protection from sickle cell

Should We Be Screening For Any Problems?

- Health problems become more similar to general population issues than to sickle cell specific issues
- Sickle Cell has variable influence on presentation and course
- Therefore routine screening and care become even more important
- Don’t consider ANY problem as just a sickle cell problem

United States Preventative Serves Task Force

- High Blood Pressure
  - But sickle cell patients have lower systolic and diastolic blood pressures compared to controls
- Lipid Disorders
  - But sickle cell patients often have low total cholesterol, triglycerides less low
- Colorectal Cancer
  - Need to Start at 40? Prostate Screening of Men at 40
- Breast Cancer
  - Need to Start at 40

United States Preventative Serves Task Force

- Depression
  - Many cultural issues
  - Chronic pain, depression, and sleep disorders commonly occur together
- Primary Prevention of Cardiovascular events
  - Especially tobacco use cessation
  - Marijuana may be just as damaging

- REGULAR SCREENING WILL ESTABLISH BASELINES TO MONITOR ANY IMPORTANT CHANGES IN ORGAN FUNCTION

Sickle Cell Specific Screening

- Heart
  - Qtc, pulmonary hypertension, hypertrophy
- Lungs
  - Restrictive disease, obstructive disease, and mixed
- Kidneys
  - Isothenuria, proteinuria
- Bones
  - Low mineralization, low vitamin D levels
- Liver
  - Gallstones, hepatic dysfunction,
- Eyes
  - Dilated retinal examination
- Vaccinations
  - For immunosuppressed people

Sickle Cell Disease Specific Yearly Tests

- HIV – Blood exposure and usually risks
- Hepatitis C – Most common Transfusion related viral infection
- Liver function – especially for siderosis
- Renal function
  - Often isothenuric
  - Proteinuria common
  - Partial distal tubular acidosis
- Eye Examination – Hemorrhages, neovascularization, ischemic areas
- ECG – for conduction changes, especially QTc
- SQUID – for any regular blood exposure
  - Check serum ferritin, iron, total iron binding capacity

Sickle Cell Disease Specific Tests every Two or More Years

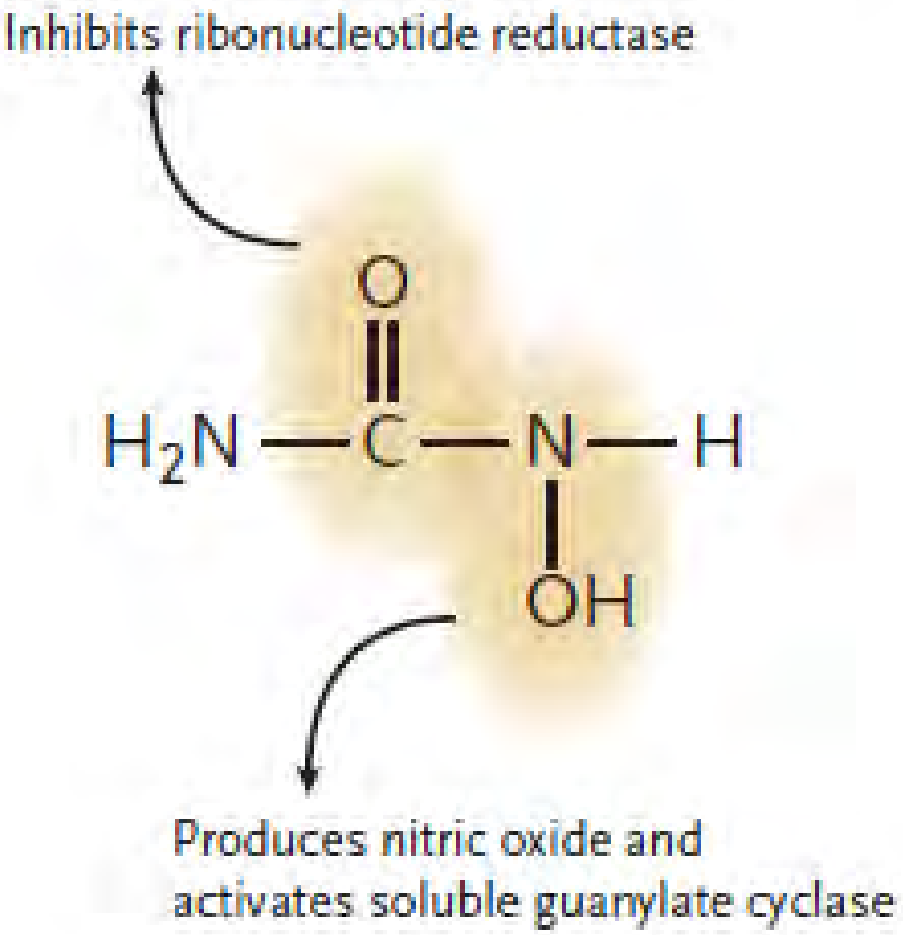
- PFTs – Often restrictive, can be severe
- MRIs – Overt and silent infarcts, volume loss, and white matter disease
- MRAs – Aneurysms, arterial narrowings, and blocks
  - Carotids?
- DXA scan – bone mineralization
- Echocardiograms for pulmonary Hypertension
  - Should be baseline with estimated tricuspid regurgitant jet (abnormal > 2.5 m/s)
  - Exercise echo with any cardiac symptoms

What Preventative Care Can Be Done For These Problems?

- Healthy Life Style
- Vaccinations Recommended for Immunocompromised Persons
- Routine lipid, diabetes, and cancer screenings
- Rapid evaluation of any new symptoms
- Consultation with the appropriate specialist
- Disease modifying treatment
- Keep medications to a minimum

Disease Modifying Treatment

- Hydroxyurea
- L-glutamine
- Transfusions
- Transplant
- Genetic Therapy



N Engl J Med 2008;358:1362-9.

Hydroxyurea in Sickle Cell Disease

- Increased fetal hemoglobin levels
- Increased hydration of erythrocytes
- Decreased adhesion of erythrocytes to vascular endothelium
- Decreased neutrophils
- Enhancement of nitric oxide

Transfusions

- How much of Symptoms in Sickle Cell are due to anemia?
- Transfusions often used for fatigue, dyspnea, and heart failure
- May protect end organs from damage
- Transfusions have improved renal and cardiac function in patients
- Little large, or long term studies of transfusions

Pain Management

- Day Hospitals reduces hospital admissions and length of stay for painful episodes
- Opioids may cause pain and prevent pain control
- Long-term effects of chronic opioids unknown, but animal studies concerning
- Monitor oxygen saturations in hospital
- Individualized Pain Plans

"Age? You mean now or when we first sat down?"

Resources

- Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014
  - http://scinfo.org/
  - https://www.nhlbi.nih.gov/health-pro/guidelines/sickle-cell-disease-guidelines/
- Sickle Cell Adult Providers Network (SCAPN)
  - www.uchsc.edu/scapn
- United States Preventative Services Task Fore
  - www.ahrq.gov/clinic/uspstfix.htm
- Centers for Disease Control
  - https://www.cdc.gov/ncbddd/sicklecell/index.html
- California State Site
  - http://casicklecell.org/