Medical Aspects of Transition in Sickle Cell Disease

Kathryn Hassell, M.D.
Professor of Medicine, Division of Hematology
Colorado Sickle Cell Treatment and Research Center
University of Colorado Denver

MMERE DANE: “time changes”
Symbol of change, life’s dynamics

Disclosures

- No financial disclosures
- No conflicts of interest

- However, an ADULT PROVIDER…

Outline of Presentation

- Medical issues for adolescents and young adults with sickle cell disease (SCD)
- Ideal adult context for SCD medical management
  - Individual's (patient's) role
  - Provider's role
- Current reality
- Potential impact of healthcare reform

Medical Issues in Adults with SCD

- Increased utilization of health care services age 18–30 years
  - Utilization
  - Readmission

Table 2. Rates of Acute Care Encounters by Age for Patients With Sickle Cell Disease

	All Patients				
	Patients, No.	Encounters per Patient per Year, No. (95% CI)			Patients, No.
		ED Visits	IP Stays	Total	
Age, y					
1-9	4251	0.59 (0.56-0.62)	0.91 (0.87-0.96)	1.50 (1.45-1.55)	3060
10-17	3605	0.68 (0.63-0.73)	1.37 (1.30-1.44)	2.04 (1.95-2.13)	2775
18-30	6377	1.59 (1.50-1.68)	2.02 (1.94-2.10)	3.61 (3.47-3.75)	5142
31-45	4540	1.29 (1.20-1.38)	1.65 (1.55-1.75)	2.95 (2.80-3.10)	3479
46-64	2040	0.86 (0.75-0.97)	1.23 (1.14-1.32)	2.09 (1.93-2.25)	1360
≥65	299	0.33 (0.13-0.53)	0.72 (0.64-0.80)	1.05 (0.83-1.27)	130
Total	21 112	1.08 (1.04-1.11)	1.52 (1.48-1.55)	2.59 (2.53-2.65)	15 946

Abbreviations: CI, confidence interval; ED, emergency department; Hb SS, hemoglobin SS; IP, inpatient.

Brousseau, *JAMA* 303:1288, 2010

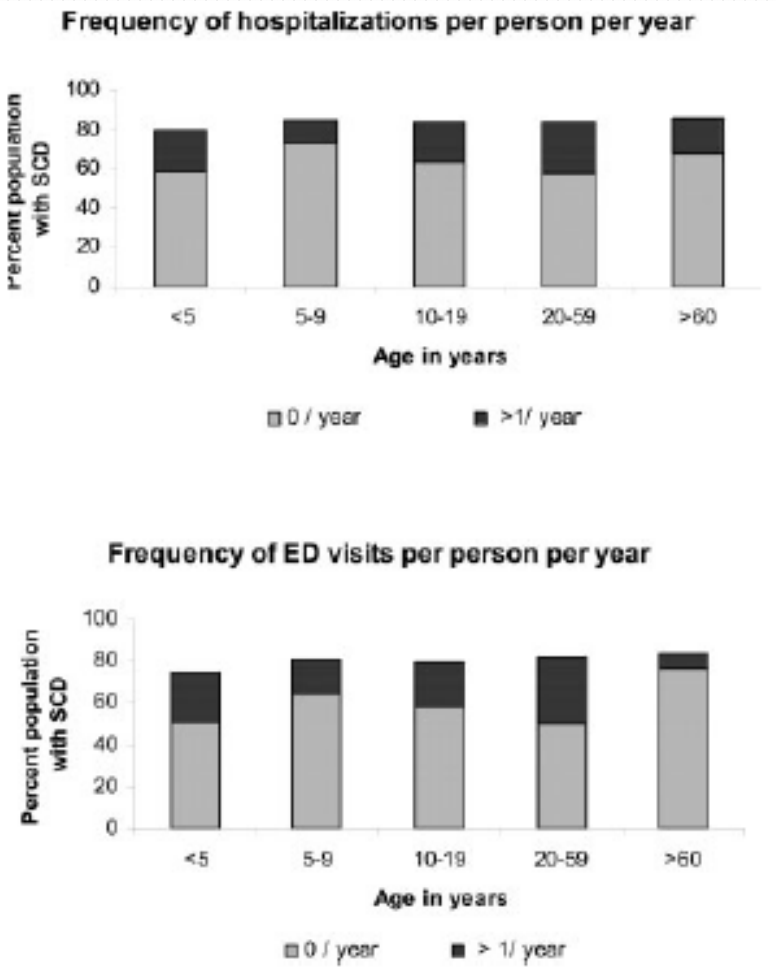


Fig. 1. Frequency of hospitalizations and ED visits in patients with SCD. ***Note:*** Only 22 individuals are in the age category >60 years, and hence the data in this category should be interpreted with caution.

Shankar, *Am J Hem* 80:262, 2005

Medical Issues in Adults with SCD

- Causes of death
  - Changing from acute to chronic conditions

Time period	Location	#1	#2	#3	#4	#5
1952-82	Jamaica	ACS (22%)	Renal failure (14%)	Infection (7%)	Pregnancy (9%)	CVA (6%)
1978-88	U.S.	ACS (14%)	Renal failure (11%)	Stroke (7%)	Peri-op (7%)	Trauma (7%)
10 yrs (<2002)	Europe	MOFS (21%)	Infection (16%)	ACS (15%)	Cirrhosis (10%)	Stroke (8%)
1976-2001*	U.S.	"PHT" (26%)	Renal failure (23%)	Infection (18%)	VTE (15%)	Cardiac (12%)

*cirrhosis – 11% ACS – 10%

Darbari, *Am J Hem* 81:858, 2006

Medical Issues in Adults with SCD

- Duke cohort, mean age 39 (dead), 34 (alive)

TABLE IV. Clinical Premorbid Complications Observed in the Study Subjects

	Deceased ($N = 43$) Number (%)	Living ($N = 196$) Number (%)	P value*
Other SCD-related complications			
Cholethiasis	24 (55.8)	115 (58.7)	0.74
Proteinuria	22 (51.2)	32 (16.3)	<0.0001
Creatinine >1.0 mg/dL	18 (41.9)	27 (13.8)	<0.0001
Avascular necrosis	16 (37.2)	13 (6.6)	<0.0001
Creatinine >1.4 mg/dL	12 (27.9)	55 (28.1)	1.00
Lower extremity ulcers	9 (20.9)	28 (14.3)	0.35
Priapism	5 (11.6)	28 (14.3)	0.81
Worsening anemia	5 (11.6)	49 (25)	0.07
Retinopathy	4 (9.3)	18 (9.2)	1.00
Seizures	3 (7)	12 (6.1)	0.74
Human immunodeficiency virus	2 (4.7)	0 (0)	0.03
Hepatitis C virus	2 (4.7)	10 (5.1)	1.00

Fitzhugh, *Am J Hem* 85:36, 2010

Medical Issues in Adults with SCD

- Duke cohort, mean age 39 (dead), 34 (alive)

TABLE IV. Clinical Premorbid Complications Observed in the Study Subjects

	Deceased ($N = 43$) Number (%)	Living ($N = 196$) Number (%)	P value*
Cardiopulmonary complications			
Acute chest syndrome/pneumonia	26 (60.5)	108 (55.1)	0.61
Pulmonary hypertension	18 (41.9)	36 (18.4)	0.001
Systemic hypertension	11 (25.6)	36 (18.4)	0.29
Congestive heart failure	11 (25.6)	16 (8.2)	0.003
Stroke	11 (25.6)	31 (15.8)	0.18
Myocardial infarction	9 (20.9)	3 (1.5)	<0.0001
Pericardial effusion	4 (9.3)	5 (2.6)	0.06
Pulmonary embolus	6 (14)	15 (7.7)	0.23
Atrial fibrillation	4 (9.3)	1 (0.5)	0.004
Supraventricular tachycardia	1 (2.3)	0 (0)	0.18
Ventricular fibrillation	1 (2.3)	0 (0)	0.18

Fitzhugh, *Am J Hem* 85:36, 2010

Medical Issues in Adults with SCD

- Duke cohort

TABLE III. Survival Age and Causes of Death for Deceased Subjects

Median survival age[a]	Years (95% CI)
Total	40 (35, 53)
Males	40 (34, 48)
Females	39 (33, 56)
Age at death (years)	Number of patients
20–29	8
30–39	15
40–49	8
50–59	7
≥60	5
Cause of death	Number of diagnosis
Cardiac	
Pulseless electrical activity arrest	5
Congestive heart failure	3
Myocardial infarction	3
Pulmonary	
Pulmonary embolus	4
Pulmonary hypertension	1
Respiratory disorder	1

Most deaths (34%) occurred ages 30-39

40% cardiopulm deaths

Fitzhugh, *Am J Hem* 85:36, 2010

Medical Issues in Adults with SCD

- Duke cohort: 32% of deaths attributed other (non-cardiopulmonary) SCD complications

TABLE III. Survival Age and Causes of Death for Deceased Subjects

Other complications of SCD	
Stroke	4
Multiorgan failure	3
Liver failure	2
Chronic renal failure	1
Anoxic brain injury	1
Sickle cell anemia	3
Other	6[b]
Unknown	6

[a] Median survival age and 95% CI are calculated from the Cox model.

[b] Foreign body aspiration (2), intestinal disorder (1), complications after fall (1), narcotic overdose (1), assault (1).

Fitzhugh, *Am J Hem* 85:36, 2010

Take-Home Messages

- Somewhere between 20 and 40 years of age, the burden of chronic organ damage creates increased morbidity and mortality
  - Cardiopulmonary disease
  - Renal disease
  - Sequelae of interventions e.g. iron overload, Hep C
- Screening tests exist to identify these conditions
  - Data are lacking as to specific interventions that may change the course of disease
- A portion of mortality is related to sudden death for which etiology and prevention are unknown

What I Do In Clinic for a Late Adolescent or Young Adult With Sickle Cell Disease

- Take a history
  - Current and chronic symptoms, e.g.
    - Pain: frequency, intensity, methods of relief
    - Cardiopulmonary: lung disease
    - RUQ/LUQ pain: gallbladder/spleen
    - Hip/shoulder pain: avascular necrosis
    - Skin breakdown
  - Medical events since last visit
    - Hospitalizations
    - Events treated at home
  - Current therapies and medications with doses
    - If on transfusion: type, total, last, on chelation?

What I Do In Clinic for a Late Adolescent or Young Adult With Sickle Cell Disease

- Take a history
  - Current/previous habits
    - Smoking, alcohol, recreational drugs
  - Social history
    - Working? In school? Sexually active? Planning pregnancy?

What I Do In Clinic for a Late Adolescent or Young Adult With Sickle Cell Disease

- Perform an examination
  - Review vital signs
    - Includes pulse oximetry
  - Lungs
  - Heart – for increased P2
  - Abdomen – for heptomegaly, splenomegaly
  - Extremities – for edema, ulcers
- Do not routinely examine:
  - Retina
  - Cervix/ovaries
  - Breasts

What I Do In Clinic for a Late Adolescent or Young Adult With Sickle Cell Disease

- Review previous/most recent evaluation
  - Clinical: ophthalmology, other specialists "prn"
  - Blood: ferritin, Hepatitis/HIV tests, RBC minor antigen testing
  - Imaging: echocardiography
  - Liver iron concentration (bx, imaging) "prn"
- Obtain and review:
  - CBC, reticulocyte count
  - Basic metabolic panel (Na^+, K^+, glucose, creatinine)
  - Hepatic panel (at least 2-3 times/year)
  - Urinalysis/microalbuminura (annually and "prn")

What I Do in Clinic for a Late Adolescent or Young Adult With Sickle Cell Disease

- Arrange follow-up to complete SCD-related evaluation as needed
- Defer other medical issues to primary care provider, e.g.
  - Contraceptive therapy
  - Pharmacological management of depression and anxiety
  - Any symptoms unrelated to SCD

Pain Management for SCD

- Majority of adults (adolescents?) experience frequent, eventually chronic, pain
  - Ask about it and address it
- Relatively small % require chronic opioid therapy, but for those who do:
  - Written management agreement
  - Informed consent
  - Monitoring for benefit, harm, adherence, diversion, misuse, addiction
  - Mitigation strategies if problems suspected

Chou, *J Pain* 10:113, 2009

What I Do When a Late Adolescent or Young Adult is Admitted

- Provide consultation to admitting physicians
- Basic management
  - Pain control: PCA or standing dose i.v. push, using individual's preferred approach and medication
  - Maintain euvolemia and normal oxygenation
  - Investigate for infection, reduce inflammation
  - Monitor vital signs carefully for tachycardia, hypoxia, fever
  - Monitor labs – CBC, creatinine
- Transfusion therapy (minor-antigen matched) for acute severe end-organ damage

Sound Familiar? It Should!

- A 19-year-old who transitioned
  - Medically speaking is the same person who just left their pediatric provider a month [or year(s)] ago
  - The medical management should be the same, regardless of who is providing it or the setting
  - Basic approaches to medical issues used by providers are basically the same regardless specifics of training
- What does the adult provider want to know about a young adult? The same things a pediatric provider needs to know if they manage individuals in this age group.

Medical Management of Adults with SCD

- Limited evidence upon which to base SCD management guidelines
- However, in the absence of randomized controlled trials, experience provides evidence:
  - Lottenberg, ASH Education Program, *Hematology*:58, 2005
  - Pack-Mabien, *J Am Acad NP* 21:250, 2009
  - Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK www.**sicklecell**society.org/pdf/CareBook.pdf
- Need to assure all aspects of health are addressed, including screening for non-SCD conditions

Consensus Statement on Transition: Essential Elements

- Identified health care professional
  - Assumes responsibility for current health care, coordination and planning
  - Attends to unique challenges of transition
  - Executed in **partnership** with:
    - Other child health care providers
    - **Adult health care providers**
    - **The young person**
    - **His or her family**
  - Assures uninterrupted, comprehensive, and accessible care **within their community**

Consensus Statement, *Pediatrics* 110:1304, 2002

Consensus Statement on Transition: Essential Elements

- Identification of core knowledge and skills for providers
  - Required to provide developmentally appropriate health care transition services
  - Made part of training and certification requirements for primary care residents and physicians in practice

- **Apply guidelines for primary and preventive care as for all adolescents and young adults**

Consensus Statement, *Pediatrics* 110:1304, 2002

Consensus Statement on Transition: Essential Elements

- Prepare and maintain **up-to-date medical summary** that is portable and accessible
- **Written** health care transition plan by age 14
  - Prepared with provider, young person and family
  - Describe at least
    - What services are needed
    - Who will provide them
    - How they will be financed
- Ensure affordable, continuous health insurance coverage
  - Should cover care coordination and transition plans

Consensus Statement, *Pediatrics* 110:1304, 2002

Providers and Transition

- 2002 Consensus Statement (AAP, AAFP, ASIM)
  - **"All adults with special health care needs deserve an adult focused primary care physician"**
  - Specifies transitioning process should begin by age 14
- Goal: by 2010, "all physicians who provide primary or subspecialty care to young people with special health care needs
  - Understand the rationale for transition from child-oriented to adult-oriented health care
  - Have the knowledge and skills to facilitate that process
  - Know if, how, and when transfer of care is indicated"

Pediatrics 110(6 Pt 2): 1304, 2002

In The Ideal…

- Medical transition results in age-appropriate SCD- and non-SCD-related care
- All services needed by adults and within the scope of practice of an internist or hematologist with sufficient knowledge of sickle cell disease
- As for most complex diseases, outcomes are likely better with increased familiarity with the condition
  - Patient- and disease-specific care plans can provide guidance for practitioners who are less familiar but willing

An Important Distinction

- What's different after transition is not the type of health care needed, it's the CONTEXT of the health care
- Expectations are different in the world of adult healthcare
  - Individuals
  - Providers

SCD Providers and Transition

- Expectations by adult providers of patients
  - Adequate knowledge of past medical history
  - Complete a screening device or questionnaire prior to being seen
  - Seen with parent or primary caretaker
  - Made arrangements for financial obligation
  - Good understanding of chronic condition
  - Good home health management skills
  - Understanding of complications of SCD
  - Working knowledge of the medical system
  - Some level of independence from parents/family

Telfair, *J Health Care Poor Underserv* 15:443, 2004

SCD Providers and Transition

- Five methods of transition
  - Ceasing to see patients with their parents
  - Encouraging patients to accept more responsibility
  - Providing literature
  - Making the patient more financially responsible
  - Having family conferences to discuss transition
- Minority providers more likely than Caucasian providers to
  - Include financial responsibility
  - Expect all methods listed (expected by twice as many providers)

Telfair, *J Health Care Poor Underserv* 15:443, 2004

Primary Care Providers and SCD Transition

- Survey of primary care physicians (pediatricians and internists)
  - 1288 respondents
    - Mean age: 49 years
    - Mean time in practice: 17 years

	Internist	Pediatrician
Treated SCD in residency	94%	99%
Treated SCD in practice	57%	70%
SCD center easily available	15%	63%
Hematologist available	89%	87%

Okumura, *J Gen Intern Med* 23:1621, 2008

Primary Care Providers and SCD Transition

- Survey results (cont'd)
  - Who would be the best primary care provider for an 18-year-old with SCD?

Preferred provider	Internists	Pediatricians
Pediatric generalist	3%	25%
Pediatric hematologists	3%	18%
Adult generalist	48%	34%
Adult hematologist	46%	17%

  - Internists more comfortable providing primary care than pediatricians for an18-year-old with SCD

Okumura, *J Gen Intern Med* 23:1621, 2008

Primary Care Providers and SCD Transition

- Survey results (cont'd)
  - Comfort level with providing primary care for young adults (aged 17–25 yrs)

	Internists	Pediatricians
Sickle cell disease	32%	35%
Chronic pain	43%	16%
Depression	54%	29%
Asthma	85%	85%
Hypertension	91%	31%

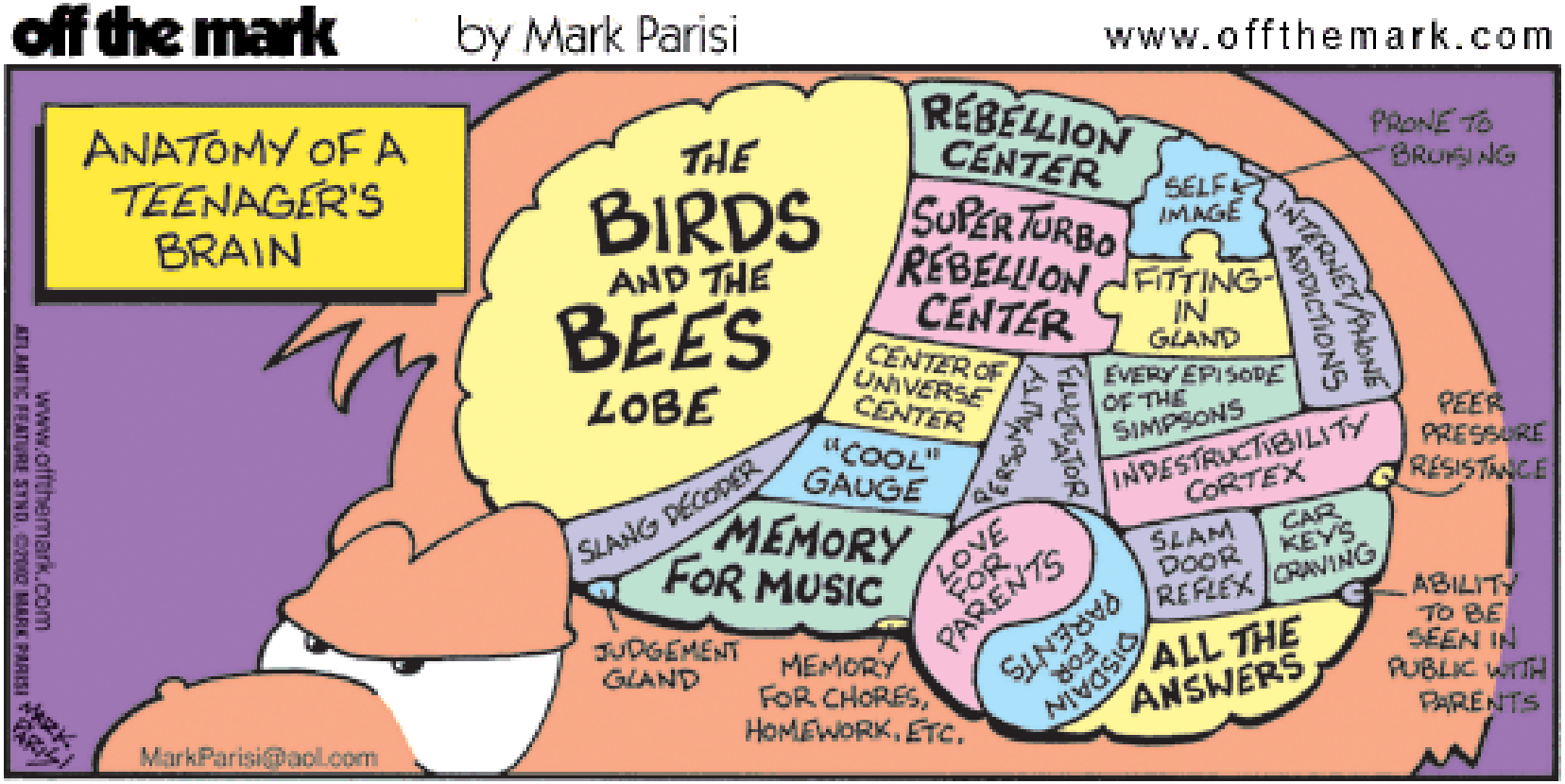
  - Comfort level with SCD was correlated with having such patients in the practice already
    - For internists only, also more comfortable if they had seen SCD patients during residency

Okumura, *J Gen Intern Med* 23:1621, 2008

Post-Transition: Context and Expectations

- Individuals are expected to behave as adults
  - Knowledge of personal health and preferences
  - Responsible for arranging care
  - Permitted to defer care, responsible for consequences
  - Responsible for financial and access issues
- (Some) adult providers, in contrast to (some) pediatric providers, expect
  - Adults are best served by adult providers
  - Report comfort with sickle cell disease if they've seen it only in residency
  - Are comfortable with issues important in adult sickle cell disease
    - Chronic organ damage, pain, general medical conditions

In Reality…



Rebellion

In Reality…

- Pediatric Provider Challenges
  - Potential for "over-protection"
    - Limiting extent of patient self-management
  - Strong bond with patients, difficult to "let go"
  - Need to develop level of trust with adult provider

    "I've been taking care of this child since he was an infant. He's doing well and I think there's some fear for really unknown reasons that I think a lot of pediatric providers feel. Like, once I transition you over to adult care, it's going to be blown"
  - Reluctance to discuss some issues due to lack of familiarity
    - Adult health care issues and complications
    - Financial/insurance considerations

Newland, *JCAPN* 21:177, 2008
Reiss, *Pediatrics* 115:112, 2005

In Reality…

- Adult Provider Challenges
  - Limited understanding/training in youth with special health care needs, including SCD
  - Potential reluctance to include parent/family
    - Although 30-60% expressed expectation to see individual with parent or family member

      Telfair, *J Health Care Poor Underserv* 15:443, 2004
  - Abrupt transfer of individuals
    - Major life events: pregnancy, "bad behavior"
    - Severe complications, chronic pain
    - No specific arrangements for insurance
  - Limited reimbursement for chronic disease management

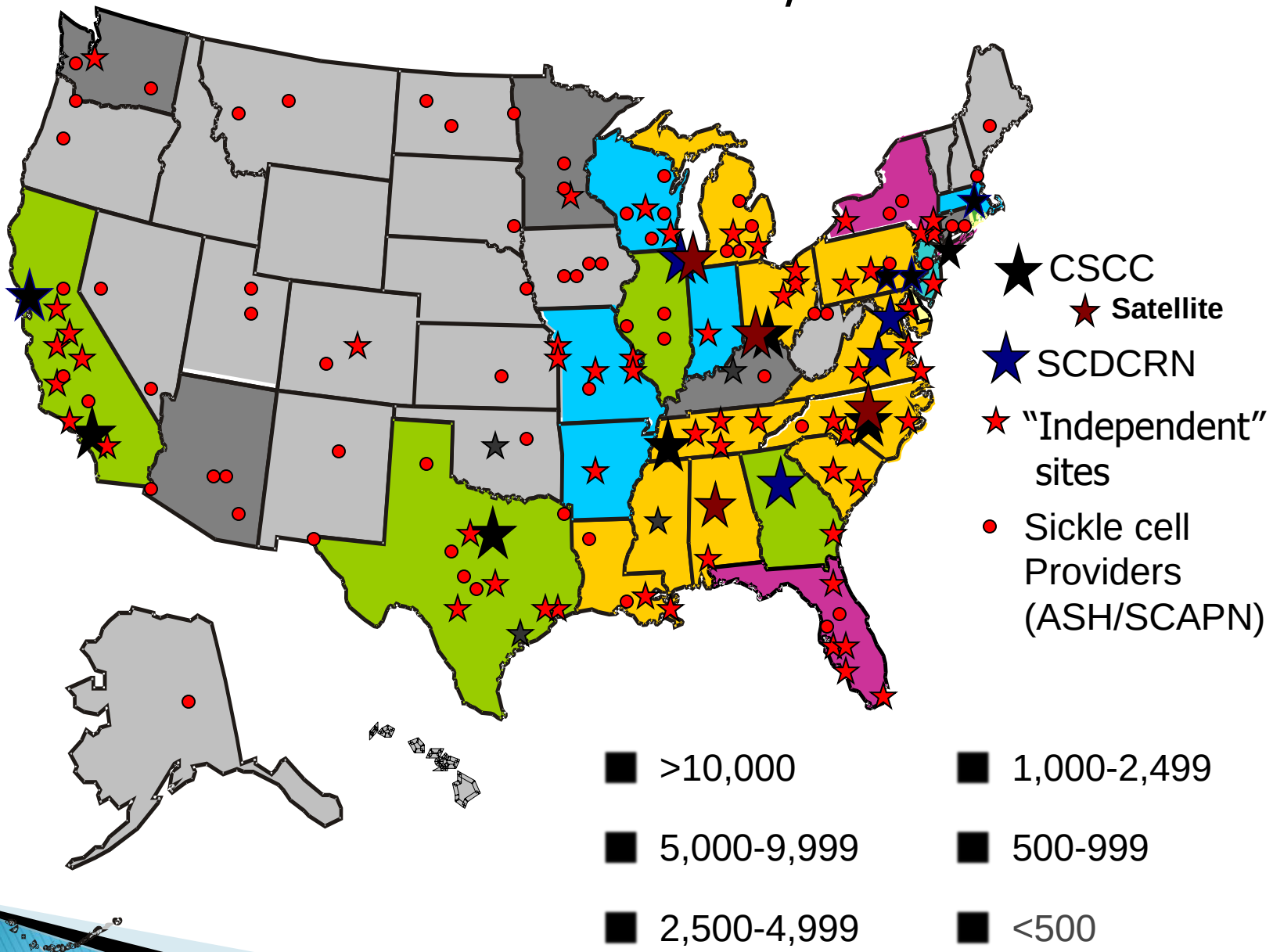
Moving Forward: Providers and Transition

- Pediatric providers must undergo transition, too
  - Expect it, plan for it, facilitate it
- Adult providers need to be a part of transition prior to transfer
- Educational/practice supports must be developed
- Providers are out there – go find them
  - Who do parents see for health care?
  - Where does the pediatric provider go for health care?
  - What clinics or providers do other adults see in the area? Do they like them?
  - What does your community support group know?

Adult Providers Exist

- Individuals with SCD in the US: ~90,000
  - Likely at least 45-50% are adults = 40,000
- At least 15,000 adults (37%) were known to academic institutions/sickle cell centers in the U.S.
  - Doesn't include those seen by primary care providers or other types of specialists

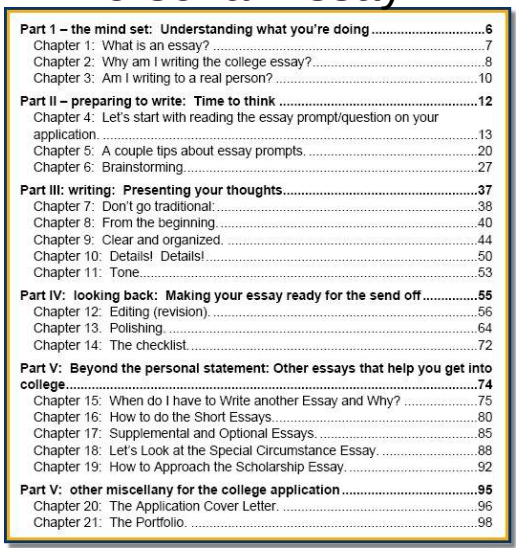
Known Sickle Cell Centers/Providers



Shouldn't Healthcare Be As Important As College?

- Applying for Higher Education

Personal Essay



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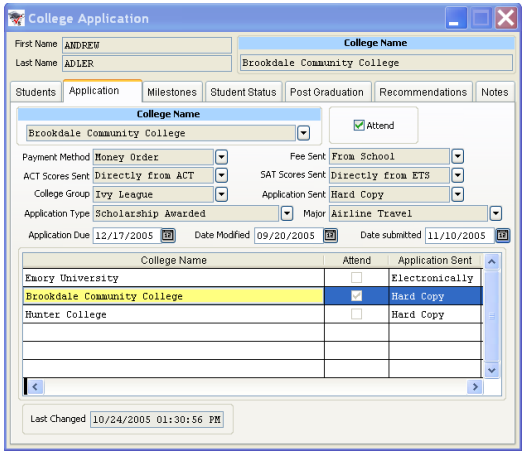
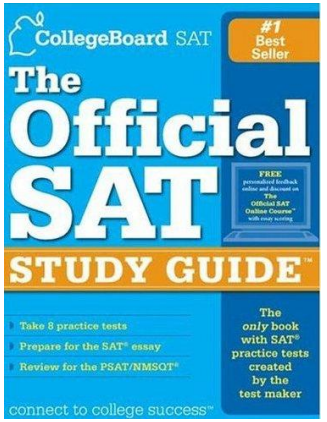


Multi-page (screen) applications



Aptitude testing

Careful review of catalogues, campus visits

PREPARATION
SELECTION

Moving Forward

- In the short run
  - Begin early to seek an adult provider and source of medical care
  - Visit several clinics/offices
  - Pediatric-adult provider contact before transfer
  - At/before transfer, send medical records
    - Send a set with the individual, too
- In the long run:
  - Medical management guidelines
  - Health care reform: adequate reimbursement for chronic disease management/medical home model