# *Best Transition Practices for Adolescents and Young Adults with SICKLE CELL DISEASE*

**EMILY RIEHM MEIER, MD, MSHS**

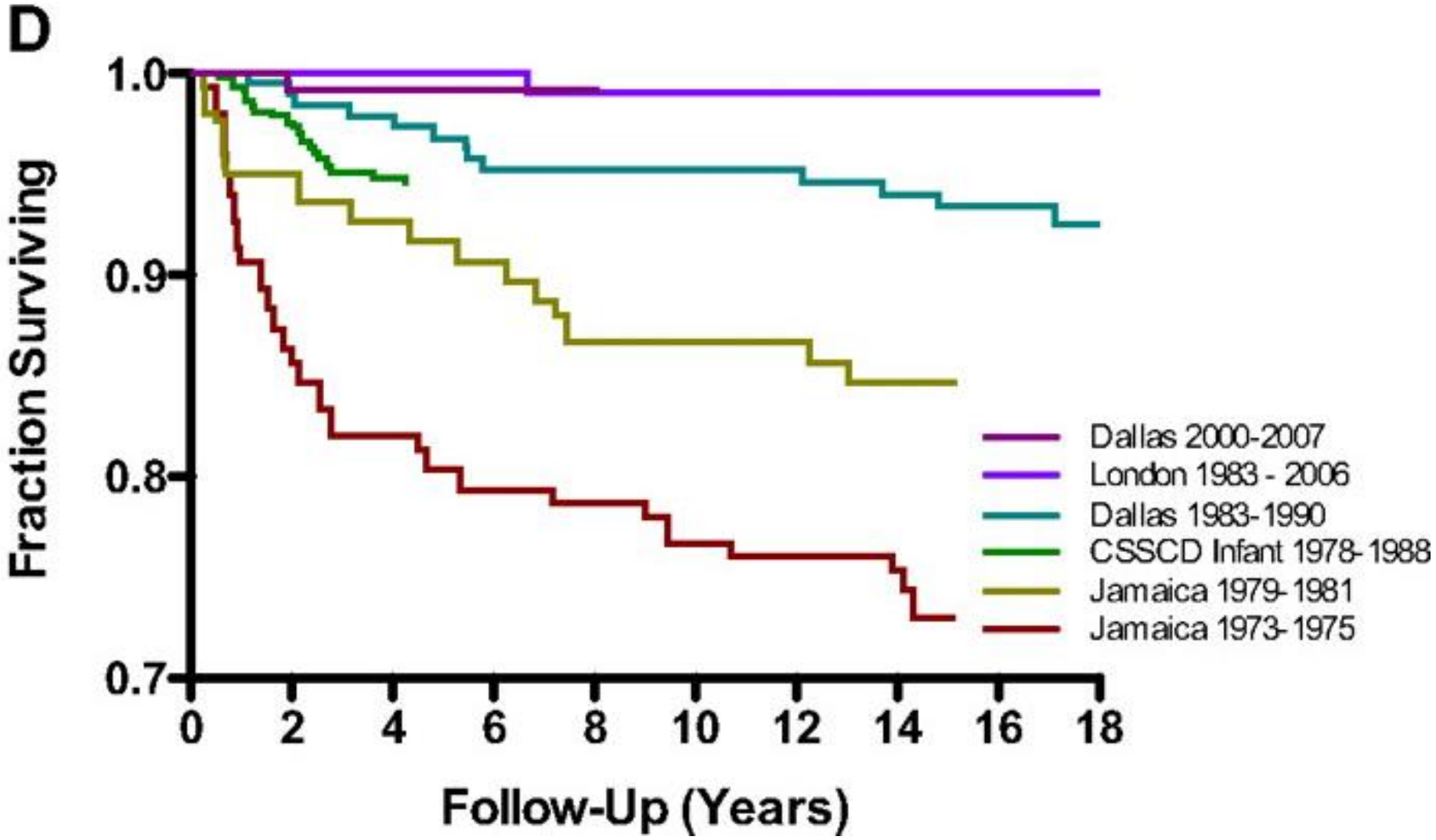
# *Disclosures*

- This Educational Module was developed through an unrestricted grant from Pfizer.

# *Objectives*

- Review emotional and social development of adolescence
- Discuss the six core elements of a transition program
- Describe the emotional and social milestones that should be addressed in a transition program for AYAs living with SCD

# *Survival of Children with Sickle Cell Disease*




*Quinn CT et al. Blood 2010;115:3447-3452*

# *Our Young Adults are at High Risk*

- The burden of mortality has shifted to young adults
- The transition from pediatric to adult medical care is a high-risk period for mortality
- Young adults urgently need the high-quality medical care and systematic follow-up that has been available to children to continue to improve the life expectancy of people with SCD.

*Quinn CT et al. Blood 2010;115:3447-3452*

# *Developmental Stages of Adolescence*

- Early Adolescence (11-13 years)
- Middle Adolescence (14-17 years)
- Late Adolescence (18-21 years)

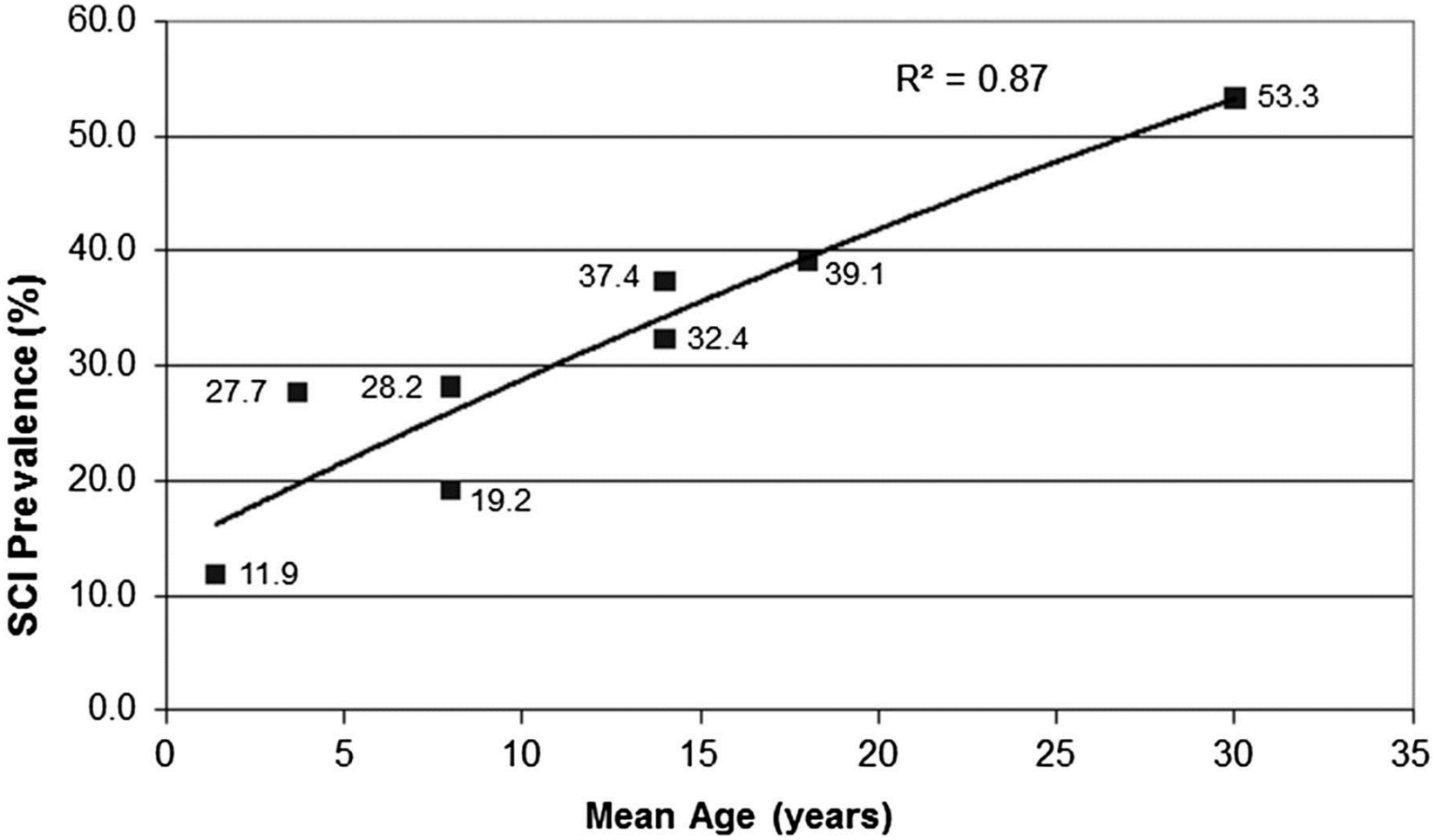
- Like infancy, cognitive, social/emotional and physical milestones should be met during these stages

| | Early Adolescence (11-13 years) | Middle Adolescence (14-17 years) | Late Adolescence (18-21 years) |
|---|---|---|---|
| Physical Growth | • Rapid growth<br>• Puberty starts | • Puberty completed | • Physical growth ends |
| Cognitive/ Intellectual | • Concrete thinking<br>• Self-centered | • Abstract thoughts<br>• Starts to set goals<br>• What's the meaning of life? | • Future oriented<br>• Philosophical<br>• Idealistic |
| Autonomy | • Challenge authority<br>• Mood swings<br>• Argumentative with parents | • Conflict with family<br>• Emerging independence | • College/work/ vocation<br>• Adult lifestyle |
| Body Image | • Preoccupied with and anxious about physical changes | • Increased interest in physical attractiveness | • Usually comfortable with physical appearance |
| Peer Group | • Intense same sex friendships<br>• Contact with opposite sex in groups | • Sexual drive<br>• Interest in dating | • Peers have less influence<br>• Selects partner |
| Identity Development | • Am I normal?<br>• Daydreaming<br>• Sexual exploration | • Experiments (sex, drugs, friends) | • Vocational goals<br>• Starts to recognize own mortality |

# *Impact of Sickle Cell on Adolescent Development*

- Physical Development
  - Delayed growth
  - Delayed pubertal development
- Cognitive Development
  - Silent Cerebral Infarcts are common
  - School absences affect education
- Social/Emotional
  - Parental overprotection
  - Uncertainty about the future
  - Denial of complications of chronic disease
  - Poor adherence to medications

# Prevalence of SCI in Children and AYAs with SCA




*Kassim AA et al. Blood 2016;127:2038-2040*



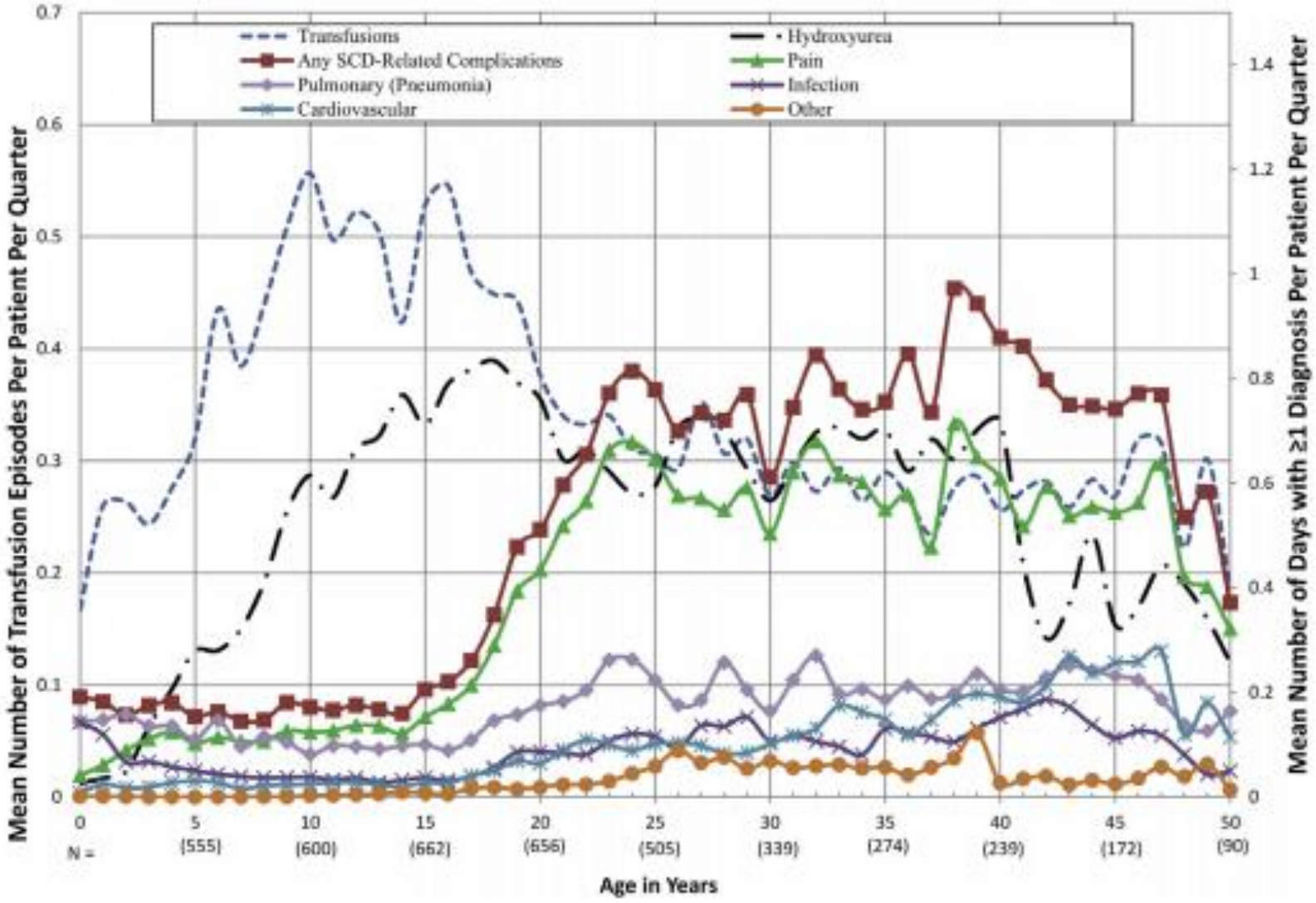
blood

# *Cognitive Impairment in Adults with SCD*

- 160 adults with SCD and 52 controls
- Neurocognitive dysfunction, undetected brain injury, or both affected most of the adults with SCD
- Areas particularly affected were those of executive functioning, reading, and mathematical ability
  - Worsening anemia associated with cognitive decline
  - 33% scored below 85 on the Wechsler Adult Intelligence Scale (WAIS) IQ scale.

*Vichinsky EP et al. JAMA 2010;303:1823-1831*

# Age-Related Care Patterns in SCD




*Blinder MA, et al. J Emerg Med 2013;49:513-522*

# *Differences between Pediatic and Adult Care*

| Pediatric Centered | Adult Centered |
| --- | --- |
| Nurturing | Informing |
| Parent Centered | Patient Centered |
| Universal funding | Unfunded |
| Family insurance provided | Employment based insurance |
| Paternalistic | Total Autonomy |
| Centralized | Fragmented |
| Informed providers | Providers unfamiliar with SCD |

# *Differences in Disease Management*

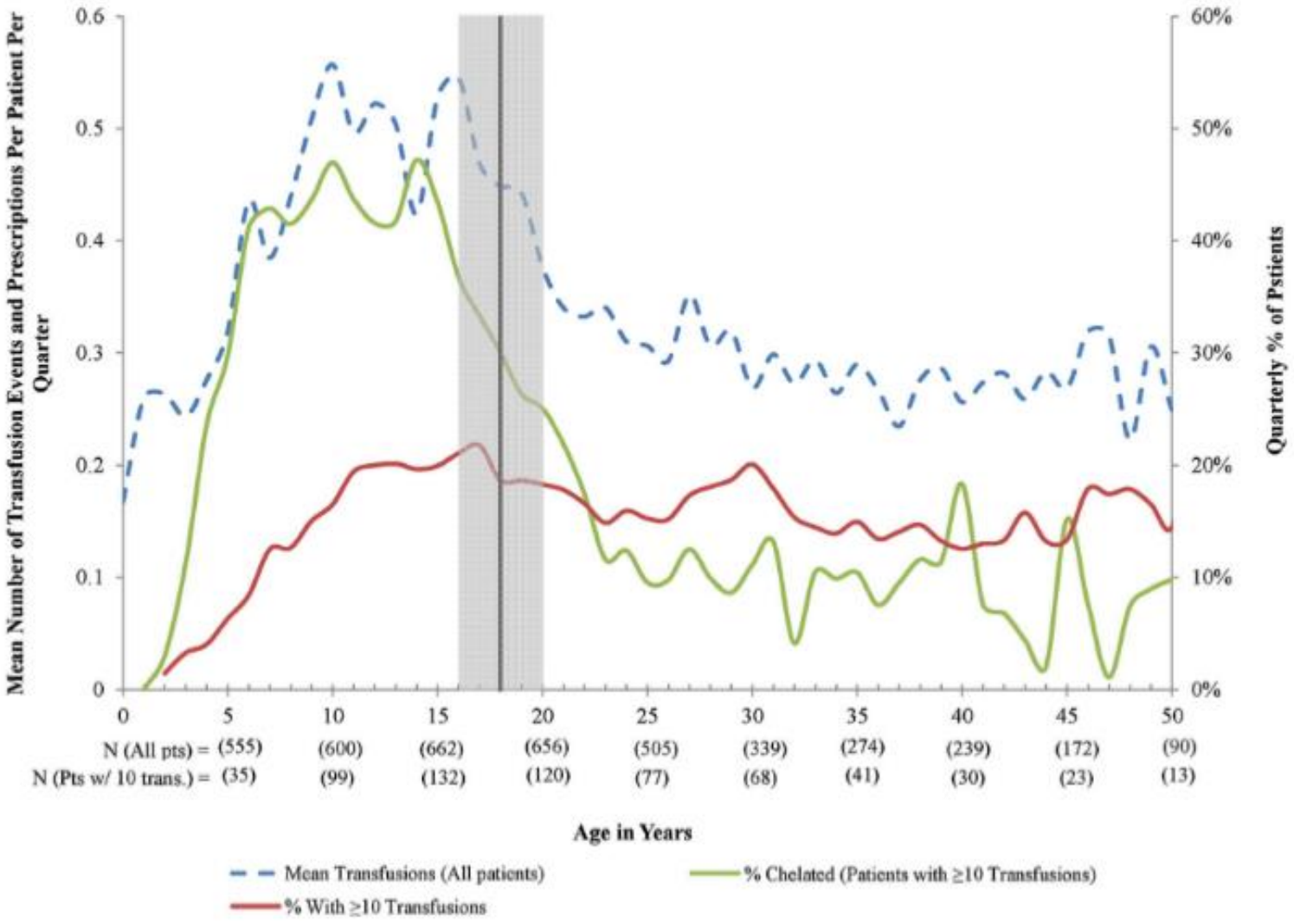
## Pediatric Sickle Cell

- Golden years
- Episodic acute pain – opioids OK
- Goal - pain free
- Decreasing problems
- Acute illness
- Immortality of youth

## Adult Sickle Cell

- Turbulent years
- Chronic unremitting pain – opioids BAD
- Goal - adjust to pain
- Progressively increasing problems
- Chronic dysfunction
- Symbiosis with disease

# Treatment Patterns by Age




*Blinder MA, et al. Pediatr Blood Cancer 2013;60:828-835*

# Age-Related Care Patterns in SCD

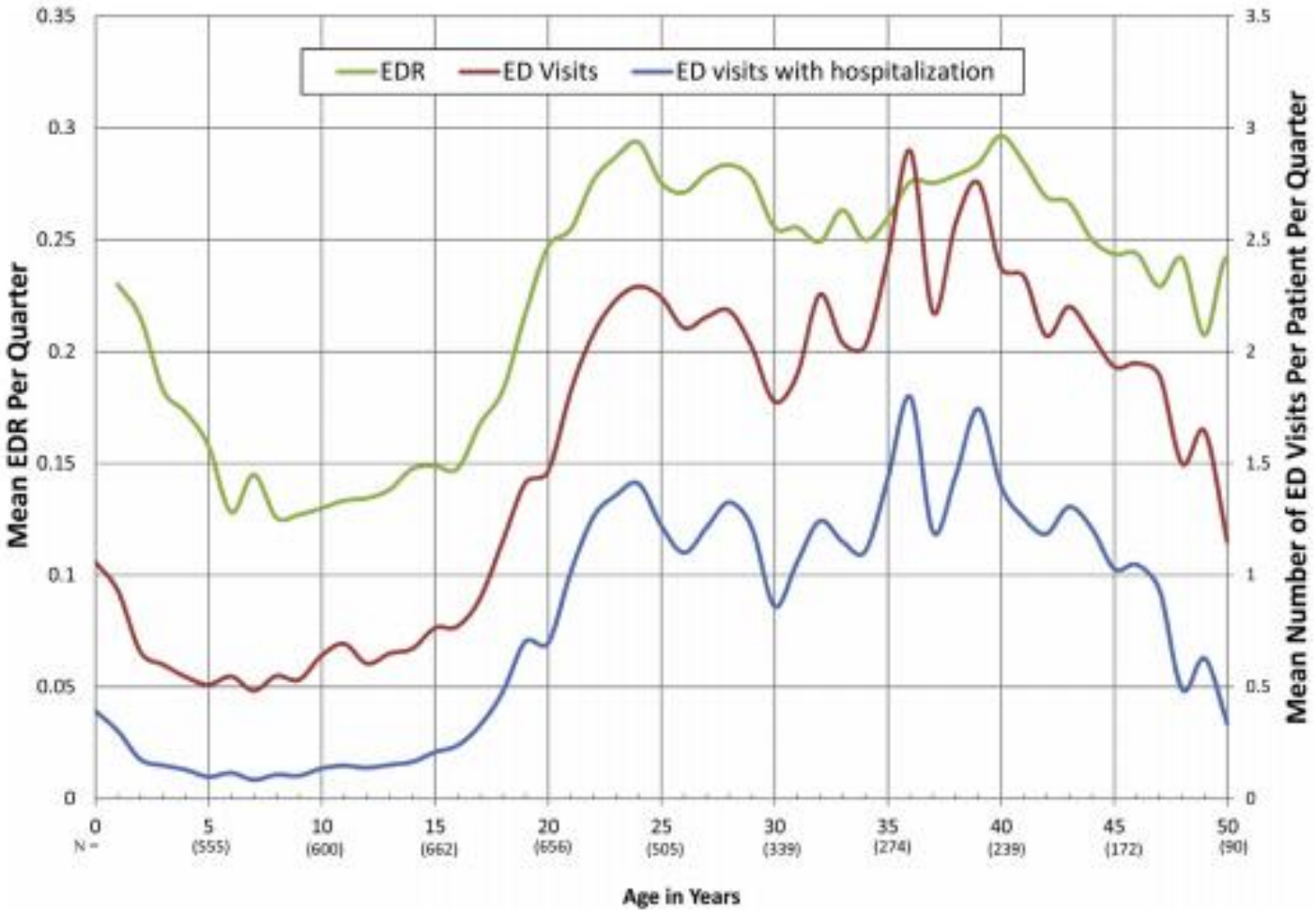
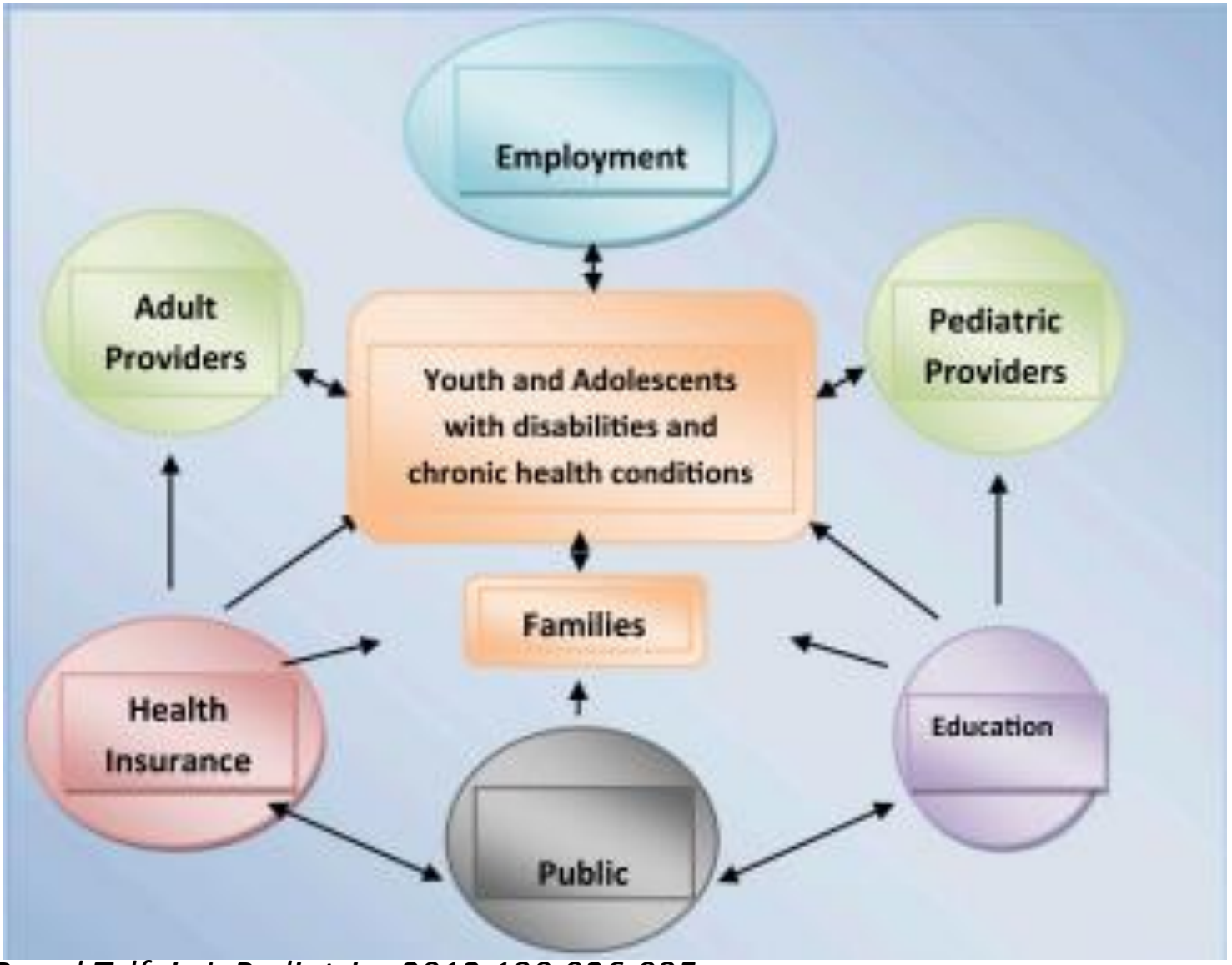



**Figure 2. Distribution of emergency department (ED) visits, ED visits requiring hospitalization, and mean Emergency Department Reliance (EDR) per quarter. *An ED visit was considered to require hospitalization if the patient was hospitalized on the same day or the day immediately after an ED visit.**

*Blinder MA, et al. J Emerg Med 2013;49:513-522*

# *Transition – An Interaction of Many Domains*




*DeBaun MR and Telfair J. Pediatrics 2012;130:926-935*

# *Recommendations to Facilitate Transition*

1) Have an orientation that is future focused.
2) Anticipate change and develop a flexible plan for the future.
3) Foster personal and medical independence and creative problem solving.
4) Anticipate future needs by developing personal life maps.
5) Celebrate transitions as they occur with graduation ceremonies, certificates of completion, and other rites of passage.

# *Six Core Elements of Health Care Transition*

- Developed by National Alliance to Advance Adolescent Health (NAAAH), using AAP/AAFP/ACP algorithm

www.GotTransition.org

- Online toolkit includes:
  - Implementation guidelines
  - Sample forms that may be edited
  - Measurement tools
- Validated in pediatric and adult academic primary care practices

*McManus M et al. J Adol Health 2015; 56: 73-78*

# AAP/AAFP/ACP Consensus Report on Transition

| | |
|---|---|
| Age 12 | • Youth and family are made aware of transition policy |
| Age 14 | • Health care transition planning is initiated |
| Age 16 | • Preparation of youth and parents for adult approach to care; discussion of preferences/timing for transfer to adult health care |
| Age 18 | • Transition to adult approach to care |
| Age 18 to 22 | • Transfer of care to adult practice |

*American Academy of Pediatrics, American Academy of Family Physicians, and American College of Physicians, Transitions Clinical Report Authoring Group. Pediatrics 2011; 128:182-200.*

# *Six Core Elements of Health Care Transition*

1. Transition Policy
2. Transition Tracking and Monitoring
3. Transition Readiness
4. Transition Planning
5. Transfer of Care
6. Transfer Completion

# *Core Element 1: Develop a Transition Policy*

- Seek youth/family/young adult input/feedback
- Entire team and medical staff need to be aware and supportive
- Post policy and share/discuss with youth and families
- Sample Policies are available at www.GotTransition.org

# *Core Element 2: Track & Monitor*

- Complete registry – see sample **Transition Registries**

- Individual transition progress should be tracked
  - Develop plan for integration of information into Health Record

# *Core Element 3: Transition Readiness Assessment*

- I know my own medicines, what they are for, and when I need to take them.
- I carry important health information with me every day.
- I understand how health care privacy changes at age 18 when legally an adult.
- I know or can find my doctor's phone number.
- I make my own doctor appointments.
- I know where to go to get medical care when the doctor's office is closed.

*Answers: "Yes, I know this." "I need to learn."*

*"Someone needs to do this…Who?"*

# *Insurance Considerations for Young Adults*

***(information current as of January 2017)***

- Stay on parent's plan
  - Young adults under the age of 26 may be eligible to be on their parent's insurance plan
- Buy an individual plan
  - Young adults >18 years of age may be eligible for savings based on income, making plans very affordable
  - "Catastrophic" health plans protect from worst-case scenarios – may be a poor alternative for persons with hemophilia
- Buy a student health plan
  - Students may be able to enroll in affordable, basic coverage through their university
- Enroll in Medicaid and/or CHIP
  - Persons with low income or certain life situations may qualify for free or low-cost coverage through Medicaid
  - Even persons who don't quality for Medicaid based on income, may have children who qualify for coverage under the Children's Health Insurance Program (CHIP)

# *Applying for SSI and SSDI*

- Supplementary Security Income (SSI)
  - 100% income sensitive – must be beneath certain income threshold to be eligible
  - Not required to have participated in the Social Security program
- Social Security Disability Income (SSDI)
  - Must have contributed to the Social Security program to participate
  - How much individual receives is based on how much individual has contributed

It takes two years to apply for Medicare once eligible individuals have been determined to be disabled, so plan ahead!

# *Self-Care Assessment for Young Adults*

- Health care provider reviews skills and knowledge required for self-care with patients over 18 years of age
- Assessment tool available on www.GotTransition.org
  - Self-Care Assessment for Young Adults
- Document in patient chart that self-care skills and knowledge were reviewed

# *Core Element 4: Transition Planning*

- Discussion of youth's/young adult's priorities and course of action that integrates health and personal goals
  - Sample Plans of Care is provided at GotTransition.org
- Prepare patient's medical summary and emergency care plan
  - GotTransition.org offers sample Medical Summaries and Emergency Care Plans
  - ASH also offers a Sickle Cell Transition Readiness Assessment and Clinical Summary in their Transition Toolkit
- Document in patient chart that transition planning was discussed

*American Society of Hematology (ASH). (no date). "ASH Releases Toolkit for Transition from Pediatric to Adult Hematologic Care." Accessed May 12, 2017, at http://www.hematology.org/Advocacy/Policy-News/2016/5581.aspx.*

# *Core Element 5: Transfer of Care*

- Transfer of Care Checklist
  - ❑ Transfer letter from Pediatrics to Adult Health Care Provider
    - ✓ Effective date of transfer of care to adult provider
    - ✓ Statement that care will be covered by pediatrics until patient is seen in adult center
    - ✓ Brief medical statement
    - ✓ Pediatrician can offer to be a consultant to Adult Health Care Provider, if necessary
  - ❑ Final transition readiness assessment
  - ❑ Plan of care, including transition goals and pending actions
  - ❑ Updated medical summary and emergency care plan
  - ❑ Guardianship or health proxy documents, if needed
  - ❑ Condition fact sheet, if needed
  - ❑ Additional provider records, if needed

# *Core Element 5: Transfer of Care*

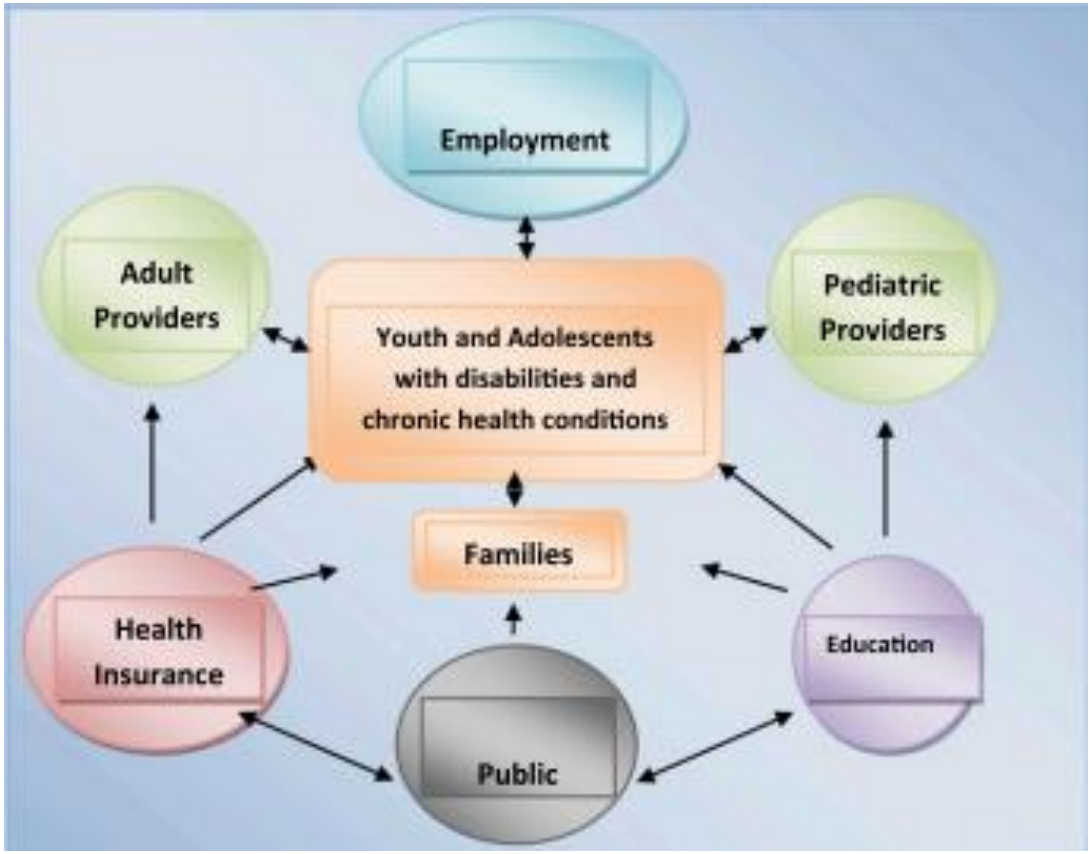
- Address any concerns that young adult has about transferring to adult approach to care
- Determine young adult's preferred mode of communication with the adult providers
- Clarify approach to adult care, including:
  - Independence in self-care
  - Adherence to care
  - Communication pathways to reach medical staff and emergency care
    - Consider health literacy, language barriers, and cultural characteristics
  - HIPAA authorization for family members and others to access health information

# *Core Element 6: Transfer Completion*

- Ask patient/family to repeat Patient Experience Survey

# *Transition and Sickle Cell Disease*

- 3 Milestones for transition to occur successfully:
  - Medical
  - Social
  - Educational




*DeBaun MR and Telfair J. Pediatrics 2012;130:926-935*

# *Medical Milestones*

- **See Module on Comprehensive care of AYAs with SCD for more information**

# *Social Milestones*

1. Health care visit with the guardian in the room but with the majority of the interview, assessment, and plan directed toward the patient
2. Instructions for medical care are accomplished initially with parental support and subsequently without parent support
3. Ability to conduct part or the entire health care visit without the guardian in the room
4. Ability to conduct a health care visit without the guardian at the health care facility, but adolescent must be of legal age
5. Ability to make appointments and follow through on self-management medical care plan without dependence on guardian
6. Visit to adult health care team office before transition
7. Ability to seek out and obtain acute medical care independently or with minimum supervision by guardian
8. Ability to determine health insurance status

# *Educational Milestones*

1. Document knowledge about sickle cell disease
2. Knowledge of current primary care health team
3. Instructions for medical care are accomplished initially with parental support and subsequently without parent support
4. Ability to describe current medications, allergies to medications, or medications that have not proven effective
5. Ability to articulate most important components of medical history and knowledge about challenges with blood transfusion therapy
6. Ability to seek out medical care with minimum parental supervision
7. Knowledge of future adult primary health care team
8. If considering offspring, awareness of partner's hemoglobinopathy trait status
9. If partner has been tested for hemoglobinopathy trait testing, counseling to assess risk of offspring having sickle cell disease

# *Summary: Best Transition Practices for AYAs with SCD*

- Adolescence has three stages and each has different developmental milestones as the adolescent grows toward independence
- Structured transition programs have 6 core elements:
  - Transition policy
  - Transition tracking and monitoring
  - Transition readiness
  - Transition planning
  - Transfer of care
  - Transfer completion
- Transition programs should allow adolescents to progressively take more responsibility for their own health care over time