Bridging the Gap: Improving Sickle Cell Disease Transition from Pediatric- to Adult-Focused Care

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Disclosures

- Advisory Board for Pfizer – Council for Change
- Cerus Advisory Board
- Novartis Advisory Board
Overview

- Discuss the challenges related to the transition of adolescents and young adults with sickle cell disease

- Describe health care utilization trends that emphasize the clinical significance around successful transition

- Outline strategies to improve the transition process for adolescents and young adults with sickle cell disease
Introduction

Sickle cell disease is complex chronic illness in which people with sickle cell disease require comprehensive care across the lifespan

- Patients, Families, Providers, Health Systems
  - Pediatric model of care
  - Adult model of care
- Community-based organization
- Federal agencies and private industry
Introduction

- Sickle cell disease is an inherited hemoglobinopathy characterized by hemolysis, vascular occlusion and unpredictable clinical complications such as acute pain, life-threatening infection, stroke and acute chest syndrome.

- Sickle cell disease has transformed from a disease of childhood to a lifelong chronic condition.

- Early diagnosis and initiation of infection prophylaxis, comprehensive care, aggressive management of acute complications, transfusion therapy for primary and secondary stroke prevention.
93% of children born with sickle cell disease will live to 18 years of age

Sickle Cell Disease: Challenges

- Continued high morbidity and mortality
- No “universal” cure
- Few disease-modifying therapies
- Poor access to high quality care
  - Need to improve existing treatment regimens
Trends in Age-Specific Death Rates in SCD 1979 - 2009

Hamideh D and Alvarez O; Pediatric Blood Cancer 2013; 60: 1482-86
Current Trends in Mortality

- When data from 1999-2009 was compared to data from 1978-1998 mortality decreased:
  - 61% in infants
  - 67% in children 1-4 years of age
  - 22-35% in children 5 years – 19 years

- After age 19 years, mortality increased from 0.6 to 1.4/100K in the 20-24 year group with a similar increase in the 1978-1998 group

- Tendency towards longer survival as more deaths occurred in 55-74 year age groups
Sickle Cell Disease: Costs of Care

Hospitalizations

- 2004 113,098 (83,149 adults) hospitalizations

December 2006

Sickle Cell Disease Patients in U.S. Hospitals, 2004

Claudia A. Steiner, M.D., M.P.H. and Jeffery L. Miller, M.D.
“For an average patient with SCD reaching age 45, total undiscounted health care costs were estimated to reach $953,640. At a 3% discount rate, the present value of lifetime costs is $460,151. Median lifetime costs were estimated at $392,940 (undiscounted) and $186,406 (discounted).”

Patient / SCD Community Challenges

- Stigma associated with acute and chronic pain management
- Challenges related to opioid prescription tracking
Sickle Cell Disease: Milestones

SCDAA established 1971

1972 Sickle Cell Prevention Act

Infection Prevention 1986

1987 NIH Newborn Screening Conference

TCD and High Risk Stroke 1992

1995 HU Therapy

1996 Stroke Prevention and Transfusion Therapy

1998 Bone Marrow Transplantation

2003 Sickle Cell Disease Treatment Act
## Sickle Cell Disease: Care Coordination Needs

<table>
<thead>
<tr>
<th>Health Care Service</th>
<th>Outpatient Visits</th>
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<tbody>
<tr>
<td>Routine Hematology</td>
<td>3 – 4 times per year</td>
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<tr>
<td>Routine Pediatric</td>
<td>1 – 3 times per year</td>
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<tr>
<td>Radiology for TCDs</td>
<td>1 – 2 times per year</td>
</tr>
<tr>
<td>Immunizations</td>
<td>At least annually</td>
</tr>
<tr>
<td>Dental</td>
<td>At least annually</td>
</tr>
<tr>
<td>Hydroxyurea Monitoring</td>
<td>At least 4 times per year</td>
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<tr>
<td>Chronic Transfusion Therapy</td>
<td>12 - 24 visits per year</td>
</tr>
<tr>
<td>Pulmonary, Cardiology, Neuro, BH</td>
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<tr>
<td>URGENT CARE</td>
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<tr>
<td>Fever</td>
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<td>Pain</td>
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Care Model

- Multidisciplinary, comprehensive care that is accessible, well-coordinated and evidence-based protocol driven
- Integrated behavioral health
- Psychosocial support in all health care settings / home
- Heath systems management
- Support for academic and vocational goals
- Integrative health component
- Community-based component
CHOP CSCC Outpatient Team

- Program Director
- Nurse Coordinator, Nurses (2)
- Program Coordinator
- Social workers (3.5)
- Behavioral Health Specialist
- Disease Educator / Education Specialist
- Community Health Worker
- Nurse Practitioners (3)
- Physicians (11 attending physicians)
- Research study coordinator
- General pediatricians, Subspecialists, Surgeons
- Ancillary care and transfusion medicine/apheresis services
- Integrative health program
- OVR/MINT Team
- Dental care
Pediatric Resources = Adult Resources?

- Physicians
- Nurse practitioners
- Social workers
- Care coordinators

- Behavioral health specialists
- Neuropsychiatric specialists
- (Transfusion medicine specialists)

- Preventive care
- Ancillary testing
- Long-term therapy monitoring
What does this mean for SCD?

- Increased numbers of AYA transitioning to adult-focused care

- Cost of care and barriers to care are high in the SCD population

- Population stigmatized by encounters with health system for pain management

- Pediatric population with increasing outpatient service needs fostering an environment of multidisciplinary, comprehensive care

- Pediatric resources are likely more accessible than on the adult care side; multiple disciplines involved in pediatric care
What makes transition for adolescents and young adults with sickle cell disease different?
What is transition?

- The purposeful, planned and timely movement from child and family-centered pediatric health care to patient-centered adult-oriented health care

- With the goal of optimizing health and maximizing each young person’s ability to attain his or her maximum potential

Society for Adolescent Medicine
Who transitions successfully?
2009-2010 National Survey of Children with Special Needs

Factors associated with transition preparation:

- Female
- Younger age
- White race
- Non-Hispanic ethnicity
- Income > 400% of Poverty
- Condition other than behavioral health
- Having a medical home
- Being privately insured

McManus et al Pediatrics 2013; 131(6):1090-7
Adolescents and young adults with SCD during “transition-age” periods are at risk for increased morbidity and mortality

- Increased hospitalization rates
- Increased length of stay for pain
Treatment Patterns by Age
Blinder et al; Pediatr Blood Cancer 2013; 60:828-35
Increased Utilization of Long-term Therapies

- Hydroxyurea is associated with decreased frequency of acute pain, acute chest syndrome and episodic transfusions also role for HU after a brief period of chronic transfusion for stroke

- Chronic transfusion therapy is associated with decreased neurologic complications, acute pain and acute chest syndrome

- A role for HU after a brief period of chronic transfusion for stroke prevention in high risk patients

- Fifteen percent of adolescents and young adults will be on chronic transfusion therapy for secondary stroke prevention

- Expert panel report proposes that we offer HU to all children with sickle cell disease (SS, SB0thal) at 9 months
Increasing patient population receiving long-term therapies such as chronic transfusion therapy and hydroxyurea therapy

Population with strokes and possibly other neurologic complications such as neurocognitive deficits

Barriers

♦ MANPOWER
  • “A major barrier for sickle cell transition programs in several cities is the ability to identify an adult provider with expertise and interest in serving this population”

♦ TRACKING OUTCOMES
  • Poor quality indicators for successful transition – lack of outcome variable associated with transition

♦ SYSTEMATIC APPROACH
  • Transition is a process, not an event
  • It is an “individualized process; “no one size fits all”
Models of Transition Programs

AAP’s Got Transition

- Establishing access to providers who are experts in the care of adults with SCD
- Establishing a skill set on disease self-management and health system navigation
- Assessing readiness for transition
- Developing objective markers of successful transition
- Innovative methods to improve communication between practitioners in pediatric and adult care
- Allowing opportunities for adolescent/young adult to interact with the adult care team
**Solutions**

- Establish systematic transition programs incorporating follow-up by pediatric institutions

- Focus on “TRANSITION²”

- Identify health care providers willing to develop expertise in the care of adults with sickle cell disease and establish links to current experts in the field

- Establish practical surrogate, objective markers for successful transition based on evidence-based NIH guidelines

- Utilize navigators and other health system workers to participate in late pediatric care and early adult care across the transition/transfer period

- Explore community-based resources to support adolescents and young adults throughout the transition process
Advocacy: Transfusion Specific

Billing codes for erythrocytapheresis in patients with sickle cell disease to treat and prevent transfusional iron overload

Apheresis services in more geographical urban and non-urban areas throughout the US

Vascular access devices appropriate for apheresis

C, E, K-matched red cell units for patients with SCD

Developing and funding additional manpower
Individual level transition goals

Assistance with identifying transfusion services post-transition

Identification of barriers to access services such as insurance or transportation issues

Maintenance of hematologic care, particularly sickle cell expertise

Maintenance of chelation therapy
Sickle cell team transition goals

- Blood bank, transfusion/apheresis and hematologic service communication and protocol development
- Communication of medical and transfusion history
- Preparation of transfer summary
Transfusion history and management plan

Indication for Chronic Transfusion Protocol
Transfusion Method
Start Date / Frequency of Transfusions
Goals of Therapy: HbS%, Hb
Type of Unit:
  Red Cell Unit Matching: C-, E-, K-matched
  Leukoreduced, Irradiated, Washed
History of Transfusion Reactions
Red Cell Antigen Phenotype, Genotype
Red Cell Alloimmunization

Communication of medical and transfusion history
Transfusion history and management plan

Premedication
Number of Units
Difficult IV Access
Complications of Procedure – Low Calcium
Labs Needed and Frequency of Labs

Challenges with Insurance and Follow-up Visits
Executive Functioning of Patient
Red Cell Transfusions for SCD

- Obtain red cell antigen phenotype prior to first transfusion
- Clarify indication
- Select method
- Select goal Hb, HbS%
- Request appropriate antigen-matched red cell unit
  - Leukocyte-reduced unit
  - Hemoglobin S negative unit
  - Irradiation
Red Cell Transfusions for SCD

- **Determine post-transfusion monitoring method**
  - Monitor red cell antibody development
  - Monitor delayed hemolytic transfusion reactions
  - Monitor iron overload
  - Monitor viral exposure
- **Patient and family education**
Summary

- Transition is difficult in populations with chronic illness but it is particularly challenging in sickle cell disease.

- Programs should have a formal transition program and identify multiple adult-focused care options for patients.

- Increased advocacy efforts are needed to increase resources for adult-focused health care services for patients with SCD.
Components of Transition for the Apheresis Team

- Disease self-management
- Navigation of health care system
- Care coordination
- Barriers to care
- Psychosocial issues that could impede successful transition
- Fostering access to adult education and adult vocational programs
Get CONNECTED

Evidence-Based Management of Sickle Cell Disease

Expert Panel Report, 2014

American Society of Hematology

STATE OF SICKLE CELL DISEASE

2016 REPORT