Disclosures

- Speaker: Novartis, Alexion

Outline

- Background and Definitions
- Medical Homes for Complex Conditions
- When
  - Perspectives of providers, parents, adolescents
- Where
  - Differences from other specialties
- How
  - Perspectives
  - Resources

Outline

SCD Model

- Challenges facing transitioning SCD patients
- Key components for a successful transition to adulthood
  - questions to ask and how to explain the answers
No Longer Just a Childhood Illness

- ~ 11.2 million children (15% of all US children) 0-17 years have special health care needs
  - 500,000 CYSHCN turn 18 and enter adulthood in the US yearly

- Survival rates have increased for children with chronic illnesses
  - >90% survive beyond their 20th birthday

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Survival Info.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Childhood Cancers</td>
<td>• 46% of survivors are 20-40 y.o.</td>
</tr>
<tr>
<td></td>
<td>• 18% of survivors are &gt; 40 y.o.</td>
</tr>
<tr>
<td>Cystic Fibrosis</td>
<td>• Median survival 37 y.o.</td>
</tr>
<tr>
<td></td>
<td>• 50% are &gt; 18 y.o.</td>
</tr>
<tr>
<td>Congenital Heart Disease</td>
<td>• 85% reach adulthood</td>
</tr>
<tr>
<td></td>
<td>• Over 1,000,000 living with CHD</td>
</tr>
<tr>
<td>Down Syndrome</td>
<td>55-year life expectancy</td>
</tr>
<tr>
<td>Hemophilia</td>
<td>60-year life expectancy</td>
</tr>
<tr>
<td>Sickle Cell Disease</td>
<td>60-year life expectancy</td>
</tr>
<tr>
<td>Spina Bifida</td>
<td>&gt; 80% reach adulthood</td>
</tr>
</tbody>
</table>

Definition of Adolescent Health Care Transition

- Transition is defined as:
  - "the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented health care systems."

- Healthcare Transition ≠ Transfer of Care
- Transition is a Process, Transfer is an Event
- Transition should begin long before the actual transfer

Simultaneous Transitions

- From pediatric child-centered care to adult oriented health services
- From living at home with family to living in the community
- From school to work

Source: Chang W. Transitioning for Chronically Ill Child & Adult Care. Aug 26, 2001


### Principles of Health Care Transition

(When)

1. A planned coordinated approach.
2. Transfer should occur at a time of disease stability.
3. Attempt for transferring and receiving teams to meet together with the patient/family.
4. Continuously encouraged to increase self-reliance and self-care, well prior to the anticipated transfer time.
5. Transition roles from direct caregivers to patient, advocates, supporters.
6. Health education for the adolescent.
7. Support the concept of transition.

---

### Pediatric Perspective

<table>
<thead>
<tr>
<th>Age at Which Pediatricians Think Transition Should Begin</th>
<th>Adolescents with Special Needs</th>
<th>Adolescents without Special Needs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ages</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;12 years</td>
<td>3%</td>
<td>2%</td>
</tr>
<tr>
<td>12-14 years</td>
<td>6</td>
<td>6</td>
</tr>
<tr>
<td>15-17 years</td>
<td>25</td>
<td>26</td>
</tr>
<tr>
<td>18-20 years</td>
<td>62</td>
<td>65</td>
</tr>
<tr>
<td>Don’t Know</td>
<td>4</td>
<td>2</td>
</tr>
</tbody>
</table>

McManus et al. 2008
### Pediatric Perspective

<table>
<thead>
<tr>
<th>Barriers Affecting the Provision of Transition Services</th>
<th>Major %</th>
<th>Rank</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lack of available adult PCPs (Family Med or Internal Med)</td>
<td>41</td>
<td>1</td>
</tr>
<tr>
<td>Lack of available adult specialists</td>
<td>40</td>
<td>1</td>
</tr>
<tr>
<td>Lack of knowledge about or linkages to community supports</td>
<td>39</td>
<td>2</td>
</tr>
<tr>
<td>Lack of insurance reimbursement for transition services</td>
<td>38</td>
<td>2</td>
</tr>
<tr>
<td>Fragmentation of primary and specialty adult health care</td>
<td>39</td>
<td>2</td>
</tr>
<tr>
<td>Lack of sufficient staff time to provide transition services</td>
<td>36</td>
<td>2/3</td>
</tr>
<tr>
<td>Lack of pediatric staff skills in transition planning</td>
<td>33</td>
<td>3</td>
</tr>
<tr>
<td>Difficulty in breaking bond between pediatrician and pts</td>
<td>32</td>
<td>3</td>
</tr>
<tr>
<td>Lack of adolescent knowledge of condition or self-care skills</td>
<td>19</td>
<td>4</td>
</tr>
</tbody>
</table>

McManus et al. 2008

### Adult Medicine Perspective

<table>
<thead>
<tr>
<th>Barriers Affecting the Transfer to Adult Providers</th>
<th>Rank</th>
</tr>
</thead>
<tbody>
<tr>
<td>Personal competence/Need for super-specialists</td>
<td>1</td>
</tr>
<tr>
<td>Not enough family involvement (especially for cerebral palsy, MR)</td>
<td>1</td>
</tr>
<tr>
<td>Families expect significant time/attention for care</td>
<td>1</td>
</tr>
<tr>
<td>End of life issues</td>
<td>1</td>
</tr>
<tr>
<td>Adolescent/young adults self-care skills and knowledge</td>
<td>2</td>
</tr>
<tr>
<td>Insurance concerns</td>
<td>2</td>
</tr>
<tr>
<td>Lack of staff skills in care for these patients</td>
<td>2</td>
</tr>
<tr>
<td>Lack of knowledge about or linkages to community supports</td>
<td>2</td>
</tr>
<tr>
<td>Inadequate transfer of medical information/records</td>
<td>2</td>
</tr>
</tbody>
</table>

Peter NG et al. 2009

### Parent Perspective

- 50% of parents reported discussing transition with their pediatrician
- 21-30% had discussed changing to an adult-oriented health provider
- 30% had a plan in place for transition

2001 National Survey of Children with Special Health Care Needs


### Stages of Transition

- **Envisioning a Future (diagnosis)**
  - At the time of diagnosis
  - Maintaining a future orientation

- **Age of Responsibility (pre-teen)**
  - Self-care skills development
  - Decision-making rights and responsibilities

- **Age of Transition (adolescence)**
  - Practicing Interdependence
  - Formal "graduation" and transfer of care

Health Care: Transition, Family, and Pediatric Perspectives. July 15, 2005, Skokie, IL. (n=1175)
Finding Adult Providers (Where)

- Experience with adults and children increases comfort level with developmental conditions (Family Medicine and Med-Peds)
- Having $\geq 1$ patient with the same diagnosis increases comfort level

Helping Families Find New Providers (Where)

- Ask family advocacy groups, MR/DD agencies, local group home provider
- Transfer while still on family private insurance
- Ask for long visit (before lunch, end of day)
- Transfer information before first visit
- Provide resources to new provider (info on diagnosis)

Adolescent Self-Management Skills (How)

- Self-care skills were identified by pediatrics, adult medicine, parents, and adolescents as critical to successful transition.
  - In a Cincinnati survey, 80% of the adult providers felt the transferred adolescents lacked adequate self-care skills
- Education
  - Functional knowledge > book knowledge
  - "Teach-back" assesses understanding
  - Family and other providers

Adolescent Self-Management Skills

- Cognitive level, learning disability, and health literacy can impact self-care abilities
  - "Typical appearing" adolescents with special health care needs often have "hidden" learning disabilities
- Testing
  - Neuropsychological ideal (difficult to obtain)
  - Language, particularly higher level testing of pragmatic/social skills
Transferring Care – The Basics

- Occurs at the time of disease stability
- Many adolescents and parents believe the process should occur usually around 18-21 (19 average)*
- 1 year process
  - Refer, have initial visit, see back, feedback, fix problems
- Help family identify adult provider at the same level of service (i.e. specialist to specialist)
  - Accepts insurance (transfer when still on parents’ insurance)
  - Will follow medical condition
  - Is located reasonably close to patient
- Provide appropriate medical summary

*Yi, M. et al. 2007

CHALLENGES FACING TRANSITIONING SCD PATIENTS

Information at Transfer of Care

- Portable medical record
  - Diagnoses
  - Medications
  - Allergies
  - Procedures
  - Important and/or most recent labs and rads
  - Equipment
  - Care providers
  - Community agencies
  - Family creates

- Clinical summary
  - Medical
  - Equipment
  - Procedures
  - Labs and Rads
  - Info on condition
  - Developmental
  - Psychosocial/Family
  - Vocational-educational
  - Community/financial resources
  - Provider creates
Recent Developments Have Greatly Improved Patient Management and Survival

Transitioning Poses Many Challenges for Patients

Challenges for Any Patient With a Chronic Condition
- Inability to effectively manage their disease
- Need for effective coping strategies
- Concerns about cost of care and insurance coverage
- Uncertainty about future as an independent adult

Challenges of Particular Concern for SCD Patients
- Increased socioeconomic challenges
- Large number of older SCD patients, placing strain on the system
- Adult “transition patients” ending up in the pediatric emergency room
- Lack of a medical home with experience and history

Acute Health Care Utilization Is Highest in Transition-Age and Young Adult Patients

Readmission Rates Also Spike After Transition

<table>
<thead>
<tr>
<th>Age range, years</th>
<th>n</th>
<th>14-day Rehospitalization, n (95% CI)</th>
<th>30-day Rehospitalization, n (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-9</td>
<td>6895</td>
<td>7.3 (6.7-7.9)</td>
<td>12.8 (12.0-13.6)</td>
</tr>
<tr>
<td>10-17</td>
<td>8825</td>
<td>14.1 (13.4-14.8)</td>
<td>23.4 (22.5-24.3)</td>
</tr>
<tr>
<td>18-30</td>
<td>24,645</td>
<td>28.4 (27.8-29.0)</td>
<td>41.1 (40.5-41.7)</td>
</tr>
<tr>
<td>31-45</td>
<td>15,118</td>
<td>26.0 (25.3-26.7)</td>
<td>39.8 (38.9-39.6)</td>
</tr>
<tr>
<td>46-64</td>
<td>5,160</td>
<td>15.3 (14.3-15.3)</td>
<td>27.1 (25.9-28.3)</td>
</tr>
<tr>
<td>&gt;65</td>
<td>427</td>
<td>6.3 (4.0-8.6)</td>
<td>11.9 (8.8-15.9)</td>
</tr>
</tbody>
</table>

* Approximately 80% of rehospitalizations were to the same hospital
  - 94% of pediatric rehospitalizations
  - 77% of adult rehospitalizations
The Transition Years Are A Vulnerable Time in Life For Many SCD Patients

- Deaths in patients 18 years of age or older in the 940-patient Dallas newborn cohort
  - There were 7 deaths between 2002 and 2007, all of which occurred in transition-age patients
  - Mean time between transition and death was 1.8 years

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age at transition, y</th>
<th>Age at death, y</th>
<th>Chronic complication of SCD</th>
<th>Circumstances of death</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>N/A*</td>
<td>18.8</td>
<td>Renal failure, dialysis</td>
<td>Found dead at home</td>
</tr>
<tr>
<td>2</td>
<td>17.9</td>
<td>19.2</td>
<td>None known</td>
<td>Acute chest syndrome</td>
</tr>
<tr>
<td>3</td>
<td>18.0</td>
<td>18.3</td>
<td>None known</td>
<td>Acute chest syndrome, refused transfusion</td>
</tr>
<tr>
<td>4</td>
<td>18.2</td>
<td>20.6</td>
<td>None known</td>
<td>Fell in hospital while pregnant; cerebral hemorrhage</td>
</tr>
<tr>
<td>5</td>
<td>18.3</td>
<td>19.3</td>
<td>None known</td>
<td>Acute chest syndrome complicated by multiorgan failure syndrome</td>
</tr>
<tr>
<td>6</td>
<td>18.4</td>
<td>23.7</td>
<td>None known</td>
<td>Presumed stroke</td>
</tr>
<tr>
<td>7</td>
<td>18.5</td>
<td>18.7</td>
<td>None known</td>
<td>Multiorgan failure syndrome</td>
</tr>
</tbody>
</table>

*Transition was delayed beyond 18 years due to chronic renal failure.

Risk factors for unsuccessful transition

- Associated with unsuccessful transition (1 adult visit):
  - Transferring to adult care ≥21 (p=0.008)
  - Distance >20 miles (p=0.01)
  - Milder phenotype - SC/Sβ0 (p=0.04)
  - Not being on chronic transfusions (p=0.02)

32% did not transfer successfully

SCD Transition Program

- Comprehensive team and policy
- Pre-defined Policies
  - Age = 18yo and graduation from high school
    - 20yo if delayed graduation
  - 1-2 years in peds if attending college
  - Education = must complete level 1 and 2 education in pediatrics
    - mastery >80% on knowledge questionnaires

SCD Educational Model to Promote Successful Transition to Adulthood
SCD Transition Program

- Components:
  - Medical Care – Adolescent/young adult Health Focus
  - Support - Case Management, Social, Psychological, volunteers/educators
  - Education - Patient, Family, Providers

Planning for the Future: Levels of Patient Understanding

<table>
<thead>
<tr>
<th>Level</th>
<th>(WHO)</th>
<th>(WHAT)</th>
<th>(WHERE/WHY)</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Know who they are</td>
<td>Know what to do in different</td>
<td>Know where they are going</td>
</tr>
<tr>
<td>II</td>
<td>13 to 14 years old</td>
<td>15 to 16 years old</td>
<td>17 to 18 years old</td>
</tr>
</tbody>
</table>

WHO

Level I of Understanding

“\(I\) am _____. and I have sickle cell type _____."

Level I Understanding

Patients Aged 13 to 14 Years

Basic points to understand
- Shape of a sickle cell
- Genetics
- Types of sickle cell
- How sickle cell causes complications
- ‘Stressors’ that cause sickling
### What causes more ‘Sickling’?

- Any stress on your body
  - Dehydration
  - Infection
  - Not breathing well
  - Temperature changes

**Body happy = Blood happy!**

### Patients Should Understand Implications of Their Lab Results

<table>
<thead>
<tr>
<th>Basic points to understand</th>
<th>Warning signs for complications</th>
<th>Types of blood cells</th>
<th>Management of pain</th>
<th>Medications taken daily</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin</td>
<td>Carries Oxygen</td>
<td>MCV</td>
<td>Reticulocyte Count</td>
<td></td>
</tr>
<tr>
<td>White Blood Cells</td>
<td>Fight infection</td>
<td>Platelets</td>
<td>Transfusions, SF, LIC</td>
<td></td>
</tr>
<tr>
<td>Platelets</td>
<td>Stop bleeding</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**MCV**, mean corpuscular volume.
What Patients Need to Know About:

<table>
<thead>
<tr>
<th>Pain management</th>
<th>Shortness of breath</th>
<th>Neurologic complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Know their medications</td>
<td>• Can be an emergency</td>
<td>• Emergency</td>
</tr>
<tr>
<td>• Have a pain action plan</td>
<td>• Risk for acute chest syndrome</td>
<td>• Weakness, slurred speech, visual changes</td>
</tr>
<tr>
<td>• Take main pain medication with anti-inflammatory</td>
<td>• Asthma increases their chances of having issues</td>
<td>• Need immediate transfusion or exchange transfusion</td>
</tr>
<tr>
<td>• Drink fluids, rest, reduce 'stress' to body</td>
<td>• May need immediate transfusion or exchange transfusion</td>
<td></td>
</tr>
<tr>
<td>• Interplay between stress, swelling, and pain</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Pain management:
- Know their medications
- Have a pain action plan
- Take main pain medication with anti-inflammatory
- Drink fluids, rest, reduce ‘stress’ to body
- Interplay between stress, swelling, and pain

Shortness of breath:
- Can be an emergency
- Risk for acute chest syndrome
- Asthma increases their chances of having issues
- May need immediate transfusion or exchange transfusion

Neurologic complications:
- Emergency
- Weakness, slurred speech, visual changes
- Need immediate transfusion or exchange transfusion

WHERE/WHY
LEVEL III of Understanding

“I am _____, and I have sickle cell type ____.
Sickle cell is when your blood looks like a banana and can cause blood to have problems going places. Stress to the body causes issues.

I know what I need to do when ____ happens with sickle cell.

I am going to ____ college/university and/or I am going to work. I know about what is bad for me and who/what are bad influences.”

Level III understanding
Patients Aged 17 to 18 Years

Basic points to understand
- Health Maintenance
- School and/or work (limits and resources)
- Insurance

Important Screenings for Health Maintenance

Level III understanding
Patients Aged 17 to 18 Years

Level I
13 to 14 years old

Level II
15 to 16 years old

Level III
17 to 18 years old

Routine dental exams
Dilated eye exam
Vaccinations
Urine microalbumin
Pulmonary screening
What Patients Need to Know About:

<table>
<thead>
<tr>
<th>School</th>
<th>Work or volunteering</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Absences should be excused if they are</td>
<td>• Patients need to make sure they and their</td>
</tr>
<tr>
<td>absent for SCD issues</td>
<td>employers know their limits</td>
</tr>
<tr>
<td>• Schools should work with them to get</td>
<td>• Patients should</td>
</tr>
<tr>
<td>missed assignments</td>
<td>– Hydrate</td>
</tr>
<tr>
<td>• Goals should be modified during illness</td>
<td>– Take breaks</td>
</tr>
<tr>
<td>or recovery</td>
<td>– Understand when they need</td>
</tr>
<tr>
<td></td>
<td>medications</td>
</tr>
</tbody>
</table>

Level I
13 to 14 years old

Level II
15 to 16 years old

Level III
17 to 18 years old

Make Sure Patients Understand Their Insurance Options

- Patients need to understand that seeking treatment without insurance will result in large medical bills
  - Leading cause for young adults to end up bankrupt is large, unexpected medical bills
- Patients should talk to a social worker or regional public health sickle cell educators about resources and options.

Coordination of Care With the Adult Provider is Crucial

- Agree upon the transfer process and time necessary for transition should be agreed upon by both
- Outline the coordination of care before and after

 Pediatric provider
- Inpatient care
- Medications
- Transfusion/exchange transfusion decisions and tracking
- Screening and monitoring
- Resources

Adult provider (hematologist)

Know your Sickle Cell

WHO:
I am____ and I have sickle cell type____.

WHAT:
I know what to do when ____ happens.

WHERE-WHY:
I know where I am going, to ____ college/ university and/or working at ____.
Future of medical care

Barriers to Adherence Among Patients with SCD

- Objective measures suggest medication adherence rates of 12-80% among pediatric with SCD
- Non-adherence may be associated with higher rates of vaso-occlusive crises and hospitalizations

Patient-Reported Barriers to Adherence

<table>
<thead>
<tr>
<th>Barrier</th>
<th>% Respondents</th>
</tr>
</thead>
<tbody>
<tr>
<td>Losing Medication</td>
<td>2%</td>
</tr>
<tr>
<td>Being Overwhelmed</td>
<td>23%</td>
</tr>
<tr>
<td>Lack of Time</td>
<td>23%</td>
</tr>
<tr>
<td>Forgetfulness</td>
<td>67%</td>
</tr>
</tbody>
</table>

Forgetfulness was the most commonly reported barrier to adherence in a patient survey.

Mobile Application Features to Aid Patient Adherence

<table>
<thead>
<tr>
<th>Patient-Preferred Application Features* (% Respondents)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medication Reminders</td>
</tr>
<tr>
<td>Text Message Reminders</td>
</tr>
<tr>
<td>SCD Education</td>
</tr>
<tr>
<td>Medication Education</td>
</tr>
<tr>
<td>Medication log</td>
</tr>
<tr>
<td>Laboratory Monitoring</td>
</tr>
<tr>
<td>Positive Feedback</td>
</tr>
<tr>
<td>Social Media</td>
</tr>
</tbody>
</table>

Investigational Smartphone Strategies to Improve Adherence

- Sickle Cell Disease Mobile Application to Record Symptoms via Technology (SMART)*
- Mobile Directly Observed Therapy (Mobile DOT)*
- Mobile Intensive Training Program (Mobile ITP)*

Among adolescent and young adult patients with SCD*

85%
WHAT RESOURCES ARE AVAILABLE TO YOU?

..and what should a transition plan look like?

Clinical Report—Supporting the Health Care Transition From Adolescence to Adulthood in the Medical Home

- Expert opinion and consensus on practice-based implementation of transition for all youth beginning in early adolescence
- Roadmap for transition and decision-making algorithms for all youth beginning at 12 y.o.
**Additional Resources May Help Patients and Providers**

**GotTransition.Org**

- “Six Core Elements” for transition, and other tools for providers and patients.

**Additional Resources on the Web**

- Sickle Cell Information Center: [https://sickleinfoemory.edu/sickle/](https://sickleinfoemory.edu/sickle/)

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**Tools Have Been Developed to Facilitate Transition Planning**

**American College of Physicians (ACP) Adult Care Transitions Initiative**

- **Disease-specific tools to help young adults transition successfully to self-care**
- **Components include**
  - Transition readiness and self-care assessments
  - Medical summary/transfer record
- The American Society for Hematology (ASH) participated in the initiative and developed a SCD-specific toolkit.

**ASH SCD Transition Toolkit**


**ASH Transition Toolkit: A Patient Self-Assessment Template to Determine Transition Readiness**

**SCD Transition Readiness Assessment Template**

- **Disease Knowledge:** Understanding the type of SCD and types of care that may be needed.
- **Medication Management:** Understanding medications, their doses, and adherence to therapy.
- **Appointments:** Ability to conduct and manage medical care independently.
- **Insurance:** Understanding of current coverage and possession of documents.
- **Privacy:** Understanding of current coverage and possession of documents.

Available on the ASH Website: [www.hematology.org](http://www.hematology.org)

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**ASH Transition Toolkit: Clinical Summary to Facilitate Communication with Adult Providers**

**SCD Clinical Summary**

- **Medical record summary for the adult provider**
  - Completed by the pediatric provider and patient
- **Indicates essential clinical information, including:**
  - Sickle cell history and genotype
  - Complications
  - Medications and surgeries
  - Common emergent presenting problems

Available on the ASH Website: [www.hematology.org](http://www.hematology.org)

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Conclusions

- Patients with chronic diseases are living longer
- Preparation is key
  - Collaboration and communication between adult and pediatric care (and family) is needed
- Maximizing self-management skills is paramount
- Insurance and work/school issues play a significant role in transition and transfer of care
- Both primary and specialty care transfer must be considered

“Optimal health care is achieved when every person at every age receives health care that is medically and developmentally appropriate.”

THANK YOU!