Growing Up with Special Health Care Needs

Charmaine Wright MD MSHP

2/27/20
Discussion

• Introduce the Center for Special Health Care Needs
• What is transition and why is it so risky?
• Spotlight on specific populations in Delaware
  – Intellectual/Developmental Disability
  – Cystic Fibrosis
  – Sickle Cell
  – Cerebral Palsy
• Medicaid 101
• Case study: Down Syndrome Consult Program and management pearls
• Future: Delaware’s mission
Center for Special Health Care Needs

• Population: vulnerable families growing up with chronic disease at watershed moments
  – many patients transition directly from Nemours
  – others arrive at any age with a childhood-acquired diagnosis

• Medical home that provides primary and subspecialty care, social work, and care coordination
Center for Special Health Care Needs

TEAM 1

Cystic Fibrosis
APRN
PT
Nutrition
Pulmonology
SW
Hemophilia
APRN
PT
Hematology
SW
Women’s Health
Gynecology
SW/RN CM
Cerebral Palsy
Rehab med
Ortho (intermittent)
Orthotist (intermittent)
Virtual Wound Care
SW. RN CM

TEAM 2

Sickle Cell
Hematology
SW/ RN CM
Urology
Urology
SW/ RN CM
Scribe (D/C with Cerner)
Down Syndrome
Pcare
Genetics (intermittent)
SW/ RN CM

Primary Care
WH and Mary Campbell
SW
APRN
Developmental Psychiatry
Psychiatry
SW/ RN CM
Gastroenterology: March 2020
How the first nine months shape the rest of your life

The new science of fetal origins

BY ANNE MURPHY PAUL
Molecular pathways linking maternal and child health

The two main components of the epigenetic code

DNA methylation
Methyl marks added to certain DNA bases repress gene activity.

Histone modification
A combination of different molecules can attach to the ‘tails’ of proteins called histones. These alter the activity of the DNA wrapped around them.
THE FIRST 1,000 DAYS
A CRUCIAL TIME FOR MOTHERS AND CHILDREN—AND THE WORLD
ROGER THUROW
author of THE LAST HUNGER SEASON
Special health care need?

- Chronic physical, developmental, behavioral, or emotional condition
- 10 million youth in U.S. age 0-17 have special health care needs
  - 9% of children under 6 years
  - 17% of youth 12-17 years
- 90% children with chronic conditions survive into adulthood
  - Prenatal care and diagnosis
  - Early intervention and support 0-5 years
  - School support
  - “Prepared and Hopeful Youth”
The good news

• Kids born with Down syndrome live twice as long as they did 20 years ago
• 1/68 children are now diagnosed with autism with evidence based interventions that help with behaviors, communication, and socialization
• Many complex conditions require multidisciplinary community and clinical teams for life
The *Dreaded* Transition

Medical Records
DO NOT RETURN TO SENDER
The process should be a guided educational and therapeutic transition rather than an administrative event.
Transition is Risky

• 750,000 youth enter an adult system that lacks the service capacity and expertise to care for them

• We lack system-wide policies

• In healthy young people, full development of executive functioning may continue until the late 20s
Challenge After a Childhood Disease: Finding a New Doctor

By Laura LeGrande

Some rural programs are addressing a growing gap in health care: helping the millions of survivors of various childhood diseases feel comfortable when they grow up. Thanks to medical advances, there are a growing number of survivors of childhood cancers as well as patients living longer with diseases like cystic fibrosis and spinal bifida.

More children have diseases like diabetes and asthma that will follow them into adulthood. Nearly 25% of children have at least one of a list of 18 chronic conditions, according to federal survey data.

Switching to adult care from children's hospitals and pediatricians' offices, which typically happens between the ages of 18 and 21, raises a range of issues, beyond medical care. Certain services and financial aid provided to children may not be available to them as adults. Because parents are no longer legal guardians, young patients have to make more of their own decisions or have formal guardianship arrangements.

Nemours/Alfred I. duPont Hospital for Children in Wilmington, Del., employs Cory Nourie as a patient-transition social work coordinator. In addition to referrals, its Transition of Care division offers help with medical self-management skills and videos and workshops on issues such as vocational opportunities and residential options. Originally started to help cancer patients, it was expanded five years ago, “because so many other kids in the hospital didn’t have any kind of system to allow them to transition safely and effectively from our care,” says program founder Rita Moyer, a pediatric oncologist.

Nemours refers many patients to adult care at Christiana Care Health System, also in Wilmington. Though they are separate organizations, they partner together for various programs including transitions in care.

Christiana has a transition care program for young adults with special health needs and clinics for survivors of pediatric cancer, cystic fibrosis, HIV and congenital heart disease. Medical residents learn to manage patients with multiple medical and social issues including communication difficulties, tubes and stents.

“Our medical care has advanced so much that patients with illness and diseases that never reached as adult provider are walking into our emergency rooms and family doctors and college medical centers, and we have to make sure we are ready to take them,” says Jennifer LeComte, medical director of the Christiana transitions program. She also chairs a new task force for the Society of General Internal Medicine, which aims to improve care and raise awareness among medical professionals.

Nonprofits and advocacy groups are also stepping in. The Endocrine Society, a group of researchers and doctors who treat diabetes and other hormone conditions, offers online tools to help young adults and their doctors navigate to adult care from pediatric. A nonprofit, the National Alliance to Advance Adolescent Health, last year received a five-year federal grant to operate a new Center for Health Care Transition Improvement. In June, it relaunched a website, GoTransition.org, with recommendations on how to start transitions planning as early as age 12.

Adrienne D’Oria, who turned 23 in January, had had more than 30 surgeries since infancy to deal with hydrocephalus, an excessive accumulation of fluid in the brain. Her condition required a surgically implanted shunt to keep the fluid from accumulating in the abdominal cavity where it can be safely absorbed into the bloodstream.

She has had complications including malfunctioning devices, infections, memory problems and extreme headache.

Since the age of 14, Ms. D’Oria has relied on her medical teams at Nemours led by neurosurgeon Jeffrey Campbell. She praises his caring bedside manner and tends to not cutting away her hair during repeated surgeries.

Ms. D’Oria and her mother, Mary D’Oria, plan to turn to the next page...
Shared work 2019

**Team 1: 653 patients**
- Cystic Fibrosis
- Hemophilia
- Women’s Health
- Cerebral Palsy

**Team 2: 460 patients**
- Sickle Cell
- Urology
- Down Syndrome

<table>
<thead>
<tr>
<th>Top Task Types</th>
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<tbody>
<tr>
<td>Care Management</td>
<td>465</td>
</tr>
<tr>
<td>Post Discharge Follow Up</td>
<td>111</td>
</tr>
<tr>
<td>Follow Up</td>
<td>60</td>
</tr>
<tr>
<td>Sick call</td>
<td>54</td>
</tr>
<tr>
<td>Medication Issue</td>
<td>30</td>
</tr>
<tr>
<td>Social Work - Initial Evaluation</td>
<td>28</td>
</tr>
</tbody>
</table>

- Care Management: 523 tasks
- Sick call: 70 tasks
- Mediation Issue: 41 tasks
- Social Work - Initial Evaluation: 41 tasks
- Follow Up: 261 tasks
- Post discharge follow up: 112 tasks

70 sick calls
What is medical and social complexity?

- Unique specialists seen in past year >=2
- Recent ED/urgent care visits >=4
- Recent hospitalizations: composite of number and length of stay (>=2 with LOS>=14, OR 1 with LOS>30 days)
- Recent no shows >=1
- Intellectual disability diagnosis
- Guardianship in place OR needed
- Psychosocial acuity
- Psychiatric meds: antipsychotics
- ADHD meds
- Technology dependence
- Non-office visit encounters >=20

Jan S, Wright CS, Rubin D. HRSA I-ACT Learning Collaborative
Acute Care Utilizers, n=333

96% Low

4%

- Bipolar
- Guardianship
- Psychosocial acuity
Renal Transplant: Creatinine after Transition

Watson et al, Pediatric Nephrology 2000
HIV transition

Hussen et al, Transitioning young adults from pediatric to adult care and the HIV continuum in Atlanta GA: a retrospective cohort study, J Int AIDS Soc. 2017 Sep

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Frequency (%) or median (interquartile range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>45 (62.5)</td>
</tr>
<tr>
<td>Transgender female</td>
<td>4 (5.5)</td>
</tr>
<tr>
<td>Female</td>
<td>22 (30.6)</td>
</tr>
<tr>
<td>Transgender male</td>
<td>1 (1.4)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Frequency (%) or median (interquartile range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Retained in second year of adult carea (two visits or more in second year after transition)</td>
<td>68b 38 (55.9)</td>
</tr>
<tr>
<td>VL suppressed after second year of adult carea</td>
<td>38 19 (50.0)</td>
</tr>
</tbody>
</table>
Poor outcomes at transition

- Medical complications
- Limitations in health and well being
- Lack of treatment and medication adherence
- Discontinuity of care
- Consumer dissatisfaction
- Higher acute care utilization
- Higher cost of care
SERVICES OFFERED UNDER AGE 21

- Referrals and assistance in scheduling physical exams
- Hearing and vision testing
- Growth and developmental assessments
- Transportation assistance to and from appointments

SERVICES OFFERED AFTER AGE 21
IDD

“One day your kid is getting what she needs. You're able to go to work because she's being looked after. The day she turns 21 - boom - the help ends.”

— Audrey Coccia, co-founder of Vision for Equality
Delaware Current State

• 1.7 million adults in the US with intellectual and/or developmental disabilities

• DE: Prevalence of disability is 12.2% among all ages; those with cognitive disability and independent living disability are the largest groups represented

• Adult patients with IDD knit together a patchwork of health care providers especially as they age
Emergency Placement

• Defined as unexpectedly needing residential long term care in a nursing home or facility

• Tools exist for the elderly to help families plan but not for those with special health care needs
Action: Focus Groups

• Identify long term care (LTC) planning activities and develop and test a LTC planning tool
• Delaware Epilepsy Foundation Annual Conference at Christiana Ammon Center, partnership with LEND program at UD
• Caregivers, siblings, patients with IDD themselves participated in afternoon focus groups
Long term care planning

• “Looming social, economic, and moral crisis”
• Focus groups at Christiana Care Fall 2018 (also Texas Baylor, Northwell Cohen, Rowan, CHOP/Penn, Cincinnati)
  – Housing
  – Legal planning
  – Identification of primary caregiver(s)
  – Financial planning
  – Day-to-day care
  – Medical management
  – Transportation

-Donaghue Foundation
Rates of Primary Care Screening in Patients with Intellectual or Developmental Disability (IDD)

Osborn et al. Access to Cancer Screening in People with Learning Disabilities in the UK: Cohort Study in the Health Improvement Network, a Primary Care Research Database.
## Practice Advisory: Cervical Cancer Screening (Update)

The Society of Gynecologic Oncology and ASCCP endorse this document.

On August 21, 2018, the U.S. Preventive Services Task Force (USPSTF) published its final recommendation statement (1) on cervical cancer screening in average-risk women (Table 1).

### Table 1. 2018 USPSTF Cervical Cancer Screening Recommendations for Average-Risk Women

<table>
<thead>
<tr>
<th>Population*</th>
<th>Recommendation</th>
<th>Recommendation Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Women aged &lt;21 years</td>
<td>No screening</td>
<td>D</td>
</tr>
<tr>
<td>Women aged 21 – 29 years</td>
<td>Cervical cytology alone every 3 years</td>
<td>A</td>
</tr>
<tr>
<td>Women aged 30 – 65 years</td>
<td>Cervical cytology alone every 3 years OR hrHPV testing(^+) alone every 5 years OR Co-testing (hrHPV testing(^+) and cervical cytology) every 5 years</td>
<td>A</td>
</tr>
<tr>
<td>Women aged &gt;65 years with adequate prior screening</td>
<td>No screening</td>
<td>D</td>
</tr>
<tr>
<td>Women who have had a hysterectomy with removal of the cervix and do not have a history of a high-grade cervical precancerous lesion or cervical cancer</td>
<td>No screening</td>
<td>D</td>
</tr>
</tbody>
</table>

Abbreviation: hrHPV: high-risk human papillomavirus.
Action: Health E Registries

- Advocating for the change in Powerchart and by insurance companies
- Especially important for our patient population who often need sedated visits to achieve speculum exams
- We will be early adopters
Rates of Primary Care Screening in Patients with Intellectual or Developmental Disability (IDD)

Osborn et al. Access to Cancer Screening in People with Learning Disabilities in the UK: Cohort Study in the Health Improvement Network, a Primary Care Research Database.
Mary Campbell Population Health: Colon Cancer Screening

- N=68
- 37 residents >=50 years (54%)
- Only 8 appropriately screened (22%)
- Action: Cologuard screening strategy
  - Sensitivities for the Cologuard are 92-94%
  - USPTF: average risk, no superior screening strategy
  - 29 screenings completed, 3 positive
  - 1 colonoscopy pursued, adenomatous polyp removed
Work

• Community integration
• Nationally, 34% of people with intellectual disabilities are employed
  – 53% are employed competitively (i.e., work alongside people without disabilities at a market-driven wage)
  – 38% in a sheltered workshop (work centers specifically for people with disabilities)
  – 9% in other settings (e.g., are self-employed).
• Of the adults with ID employed in a competitive setting, over half (62%) have been at their job for three years or more.
• However, only 26% of employed adults with ID have full-time jobs. Only a third were offered health insurance by their employer.
• Delaware has buy in!
Cystic Fibrosis

• Chronic lung disease with high mortality, patients rely on caregivers for support throughout the lifespan
• Advance care planning often occurs late in the course of illness and advance directives tend to be documented very near end of life, if at all
• There are high emotional and physical symptom burdens associated with CF
• Symptoms are not always well managed
• Do individuals with CF and their caregivers want their CF providers to initiate serious illness conversations?
• Are adults with CF receptive to palliative care consultation?

Volunteer Action

- 40% thinking “often” of lifespan limitations
- 60% would be willing to undergo lung transplantation→scripted communication strategy may be helpful here including PCP’s and CF team
- 80% want a heads up as to when end of life issues would be discussed→telling patients when palliative/supportive care is present /will be discussed seems to be what is largely wanted
- 80% wanted the CF team to bring it up, 100% wanted it during CF appointments→Great!!
- 40% had completed health proxy forms→this can be an area of focus
- All patients were comfortable with talking about it!
Sickle Cell Disease

- Autosomal recessive hemoglobinopathy causing abnormally shaped red blood cells
- Causes microvascular and macrovascular complications
- Estimated 400 adults with sickle cell disease in Delaware
- United States: estimated 100,000 people living with SCD
- CDC: (1989-1993) 750,000 hospitalizations cost $475M
FY 18 Total Inpatient Charges by MSDRG, 45 patients

>$5 million in charges
Sickle Cell Healthcare Utilization Trends

Figures A and B show the hospital admissions and length of stay for Acute Chest Syndrome across different age groups. Figure C and D illustrate the same data for Vaso-occlusive crisis.

Sickle Cell Disease- Readmission Risk Factors

![Graph showing the probability of readmission vs. hospitalizations for SCD in the prior year for patients with and without a primary care physician.](image)

Marcella Livingston reviewed Christiana Care Health System — Aug 26

Sickle cell patients we are tired of being discriminated against here at Christiana Hospital. Patients Relations has not responded nor Diversity outreach.

Stephanie Guarino We recently opened the Sickle Cell Program at the Center for Special Health Care Needs. We’re taking new patients and I look forward to partnering with patients and families to improve care for those with sickle cell disease. (302) 320-6300 to schedule an appointment.
# Sickle Cell Team

<table>
<thead>
<tr>
<th>Service</th>
<th>Team</th>
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</thead>
<tbody>
<tr>
<td>Hematology</td>
<td>Guarino</td>
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<tr>
<td>Social Work</td>
<td>Lauren Gillespie</td>
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<tr>
<td>Primary Care</td>
<td>Wright/Petrides/Eldridge</td>
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<td>Behavioral Health</td>
<td>Wilmington team</td>
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<tr>
<td>Pain management</td>
<td>Pharmacy, palliative care</td>
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<tr>
<td>Infusion Center</td>
<td>Wilmington/Christiana</td>
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<tr>
<td>Emergency Room</td>
<td>Wilmington/Christiana</td>
</tr>
<tr>
<td>Inpatient</td>
<td>Med-peds team</td>
</tr>
</tbody>
</table>
Sickle Cell Initiatives

- C: Multidisciplinary Clinic Day
- A: Sickle Cell Community Day Sept. 19, 2018 (planned 9/20)
- R: ACCEL Utilization review, experience of pain
Experience of Pain

• INBRE scholar
• Peer support is a novel and under-studied approach to the management of chronic pain
• Pain management programs with peer support have been shown to yield positive outcomes, including an improvement in perceived quality of life, functional capacity, number of complaints about pain, and belief in pain
• In depth interviews, n=20 patients
• Analyzing now, NVIVO
Cerebral Palsy

• Permanent disorder of movement and posture
• Follows a fetal or infant disturbance
• Associated intellectual disability, seizure disorder, behavioral problems, swallowing and GI motility problems, bone disorders, and visual or hearing deficits
Type of cerebral palsy

Regional involvement
- Spastic
  - Hemiplegia
  - Diplegia
  - Quadriplegia

Global (total body) involvement
- Dyskinetic
  - Athetoid
- Ataxia
  - Dystonic
  - Ataxic

Types of cerebral palsy and areas of brain damage involved

- Pyramidal
- Extrapyramidal

- Normal
- Mild involvement
- Severe involvement

https://goo.gl/images/bBvwXu
Center of Excellence

• Nemours
• 3000 patients annually
• Gait lab, clinical trials, 5 surgeons, 3 rehab medicine doctors, and multidisciplinary same day care
• Discharge age
Stakeholder Commitment

https://www.youtube.com/watch?v=ZsUmMA3fQN4
## Cerebral Palsy Team

<table>
<thead>
<tr>
<th>Department</th>
<th>CCHS</th>
<th>Nemours partnership</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rehab medicine</td>
<td>Guest</td>
<td>Owens/McManus</td>
</tr>
<tr>
<td>Orthotist</td>
<td>Michalowski</td>
<td></td>
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<tr>
<td>Wound care</td>
<td>Telemed initiative/Prichard</td>
<td></td>
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<tr>
<td>Nurse navigator</td>
<td>Godek/RN</td>
<td></td>
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<tr>
<td>PT</td>
<td>CCHS/YMCA</td>
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<tr>
<td>Orthopedics</td>
<td>Miller/Dabney/Shrader</td>
<td>Miller/Dabney/Shrader</td>
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<td>Neurosurgery</td>
<td>Eppley</td>
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<tr>
<td>Radiology</td>
<td>Wilmington XRAY/DEXA,</td>
<td>Kecskemethy/Bacharach</td>
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<tr>
<td></td>
<td>Manzone</td>
<td></td>
</tr>
<tr>
<td>Research</td>
<td>VI</td>
<td>Swank Neuromuscular Database</td>
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</table>
Medicaid

• Disability program
• >2/3 of the more than 400 billion dollar program spent annually
• 9 million people supported
Healthcare=

- Nursing home and institutional services
- ‘81 waivers → community living
  - prevocational services: IEP → IPE
  - supported employment services
  - entitled to live in a nursing home, but NOT entitled to the rest....
  - waiting list can be long....
Home and Community Based Services and Supplies

- Case management, personal care, home delivered meals, day services, nursing services, behavioral health, supported employment and training
- Averaging $18,000 per person annually, but varied across states
A day in the life of Ridley

With Smith’s oversight, Ridley prepares meals and cleans up afterward.

Ridley and Smith share a funny moment in the kitchen of Ridley’s residence.

Each evening, Smith wraps Ridley’s leg in a pressure device that aids with blood circulation.

Smith helps Ridley check his bank balance before they go shopping.

MICHAEL BRYANT / Staff Photographer
Community Case Study: EPIC

EPIC Has Evolved To a Singular Purpose,

LIVING A SELF-DETERMINED LIFE IN THE COMMUNITY

People with disabilities have for many years belonged to the “system” rather than to the community, needing special services and places. Many of these “services” are now unnecessary given the proper supports. There are still walls separating people with disabilities from the rest of the community. EPIC seeks to break down the walls.

Adaptive Fitness

Barbara Monaghan
Bear-Glasgow Family Y Member

Here is what we do

WE MAKE YOU A PART OF THE COMMUNITY

EPIC specializes in personalized health and wellness activities and community participation for individuals with intellectual/developmental disabilities (I/DD). We support inclusive self-determined health and wellness activities of the client’s choice at the Bear YMCA.
Jessica’s story
Ambulation in all ways

- 33% in power wheelchair
- 60% in manual wheelchair
- 53% with bracing needs
Wraparound services

surround multi-problem youngsters and families with services rather than institutional walls, and customize those services.

• Process NOT service
• Strengths come first
• Power sharing, collaboration keys to success: patient, family, community resource people, professionals
• Creativity follows
Patient Case Studies
Down Syndrome 101

• Person first language
• Cognitive or intellectual disability, not mental retardation
• Incidence is 1/750-800 live births
• Duplication of the 21st chromosome

National Down Syndrome Society
You get a call. . .

- 28 yo female – working close to full time at a local business for 6 years
  - Increase in self talk
  - Performance has dropped off at work and mother is afraid she will get fired
Wait, self talk?

• Bottom line
  – Self talk is ok
  – Dementia is a very unlikely cause of functional decline in a 28 year old

• Think about differential. . . .
More about DS

• Mental health issues
  – Depression, anxiety, OCD
  – Self talk is probably a good thing!
    • It should not land you on an anti-psychotic!!

• Intellectual disability
  – Specific learning strengths/weaknesses

• Life span
  – Increased age-specific mortality rates
  – Life expectancy has increased from 9 years in 1933 to the 60s
In this case. . .

- Cerumen removal and evaluation for hearing loss
- Sleep study and initiation of BiPAP
- Screen for hypothyroidism
- Exercise regimen to help with stress and weight loss
- Social work team intervened to help out with FMLA process and return to work
Twins with autism

• 25 yo ex 29 weekers
• Mother self-directed attendant, health limitations, caregiver support
• Primary care
• Point of Hope Day Program
• Meals on Wheels
• Speech therapy ? Need for AAC device
• Skin infection and underpads
• Need a roof
32 yo with cerebral palsy

- Amenorrhea and new hyperprolactinemia
- Women’s health: sedated pelvic exam (plus MRI and labs)
- Baclofen pump
- Guardianship versus supported decision making
Center for Special Health Care Needs

TEAM 1

**Cystic Fibrosis**
- APRN
- PT
- Nutrition
- Pulmonology
- SW

**Hemophilia**
- APRN
- PT
- Hematology
- SW

**Women’s Health**
- Gynecology
- SW/RN CM

**Cerebral Palsy**
- Rehab med
- Ortho (intermittent)
- Orthotist (intermittent)
- Virtual Wound Care
- SW. RN CM

**Primary Care**
- WH and Mary Campbell
- SW
- APRN

**Developmental Psychiatry**
- Psychiatry
- SW/ RN CM

TEAM 2

**Sickle Cell**
- Hematology
- SW/ RN CM

**Urology**
- Urology
- SW/ RN CM
- Scribe (D/C with Cerner)

**Down Syndrome**
- Pcare
- Genetics (intermittent)
- SW/ RN CM

**Gastroenterology: March 2020**