Somewhere Over The Rainbow: Adult Care Model

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Gillette Lifetime Clinic Nursing Supervisor
Objectives

• Describe a workable model to provide care to older teens and adults living with Duchenne muscular dystrophy
• Identify resources that assist in the care of adult patients
• Discuss issues related to aging with Duchenne
Gillette Children’s Specialty Healthcare & Lifetime Clinic

- Gillette is a nationally & internationally recognized leader in diagnosis & treatment with childhood onset cognitive and behavioral disabilities for over 120 years
- Lifetime Specialty Care Clinic provides specialty outpatient services to age 16 & older
- Gillette Lifetime provides a coordinated clinic for adults with neuromuscular disorders
- Clinic sees approximately 120 patients with muscular dystrophy
Why an Adult Model of Care

• “Emerging adults have poorer health than adolescents or adults who are in their late 20’s”
  (Young & Calloway, 2015)

• IOM 2014 report: transition from pediatric to adult health care as a key issue to improve the health of young adults
  (White & Ardoin, 2016)
  – eighteen million emerging adults aged 18-21 suffer a chronic health condition
  – 42% reported having a lapse of care greater than 3 years

• Increased health care costs when not connected to adult primary care with significantly higher emergency department visits
  (Bonnie et al., 2015)
The Gillette Experience
Getting it done: Mobilize stakeholders

If you don’t put resources to it, it doesn’t get done!
Why a shift in the organization of health care for adults?

- Pediatric environment for adults is unsatisfactory for patient and provider
- Emphasis on self-advocacy
- Person-centered; self-directed care
- Disability with premature aging
- Lack of coordinated care
- Lack of primary care
Top Transition Concerns for Teens & Adults seen in our clinic

<table>
<thead>
<tr>
<th>Patient Perspective</th>
<th>Parent Perspective</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Medical Issues</td>
<td>1. Insurance</td>
</tr>
<tr>
<td>2. College/Jobs/ Careers</td>
<td>2. Medical Issues</td>
</tr>
<tr>
<td>3. Transportation</td>
<td>3. Self-Advocacy</td>
</tr>
<tr>
<td>5. Social Issues</td>
<td>5. Sexuality</td>
</tr>
<tr>
<td>7. Insurance</td>
<td></td>
</tr>
</tbody>
</table>
Gillette Lifetime Specialty Clinic for Adolescents and Adults

Established in 2002
Excellence in Duchenne

Certified Duchenne Care Centers

Gillette Children’s Specialty Healthcare Neuromuscular Clinic
200 University Avenue East
Saint Paul, MN 55101

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Gillette Lifetime Care Model

• Multidisciplinary Team
• Adult approach means engaging in medical decision making to best of one’s ability
• Longer visits
• Wholistic approach
• Respect for patient’s priorities
Gillette Lifetime Care Model

- Private time with patient
- Team talks to one another
- Minimizes fragmentation of care
- Promotes continuity of care
- Specialty care vs. Primary Care
Lifetime Clinic Core Outcomes

• 1. Minimize unplanned hospitalizations
• 2. Maximize functional ability
• 3. Minimize pain or distress
• 4. Maximize well being
Gillette Lifetime Care Model

* Neuromuscular Clinic Coordinator – nurse with multidisciplinary role
* Four specialty medical providers in 1 day, 1 location, 1 clinic room, 3 days per month
  + Neurology, PM&R, Cardiology, & Pulmonary
* Nurses are assigned 1-2 patients – improves quality & continuity of care
* Transition clinic with nurse practitioner
* Ancillary support services available on site for immediate consults
* On site diagnostic testing
* Patient-reported Outcome measures
* MDA Family Support & Clinic Care Coordinator
* Monthly neuromuscular education for staff

What Makes our Adult Coordinated Clinic Unique?

Clinic Visit at Lifetime

Patient Care Team Appt precedes Physician Appt.
Typical Clinic Experience

- Previsit planning with nurse coordinator
  - 3-4 hour appointment
  - 4-6 patients
  - Bring food & activities
- Registration
  - Cerner Portal sign-up
  - Questionnaires
  - MDA Care Coordinator
- Intake
  - Vital signs, height/wt
  - Screening tools
- Respiratory Therapist
  - EKG
  - PFT & ETCO2
- RN prepares information
- Neurologist
  - Genetic counselor
  - Dietitian
- Physiatrist (PM&R)
  - Therapy screening
  - Wheel chair/seating
  - Orthotist
- Pulmonologist
  - Respiratory Control Plan
- Cardiologist
  - ECHO prior to visit
- Social worker
- RN collates plan & provides patient/caregiver education
- Nurse coordinator arranges referrals & follow-up
Interdisciplinary Management of DMD

Other considerations:
- Endocrinologist
- Gastroenterologist
- Urologist
- Ear Nose and Throat
- Palliative Care
- Adult Primary Care
- Dental
- Orthodontics

*CDC Guidelines offer lifelong recommendations

(Bushby et al., 2009)
Building Partnerships in Care

- Emergency care
- Hospitalization for adults >25 yrs
- Dedicated staff
- Provider privileges
- Consulting NM team
- Staff training
- Culture of caring
Need Knowledgeable Guidance About

- Health issues
- Sexuality
- Palliative Care
- Education
- Neuropsychology testing
- Independent Living
- Employment
- Alcohol & recreational drugs
- Medical Cannabis

If not prepared in these areas, we send the message “not important” and they are unprepared to navigate the adult world.
Sexuality Resources


 Favorites
Medical Decision-Making

- Making decisions about own care
- Tools - Five Wishes
  - [https://agingwithdignity.org/shop/product-details/five-wishes](https://agingwithdignity.org/shop/product-details/five-wishes)
  - $1 each with 25 minimum order
Evidence-Based Care

Evidence alone is never sufficient to make a clinical decision. Health care providers must balance benefits and risks, inconvenience, and costs as well as the patient’s values.

Kathleen R. Stevens, 2004
www.acestar.uthscsa.edu
Outcome Measures Framework

Tier 1:
- Immunization review: influenza
- Pulmonology health – PFT trend FVC
- Canadian Occupational Performance measure (COPM)
- Patient Activation Measure (PAM)
- WHOQOL-BREF

Tier 2:
- # hospitalization in past 12 months with respiratory illness

Tier 3:
- # Diabetes screening/diagnosis
- Presence of spinal deformity and contractures
- BMI <18 or >29
- Depression screening (PHQ-2 or 9)

(Porter, 2010)
# DMD care guideline outcome measures

<table>
<thead>
<tr>
<th>Ages 0-3</th>
<th>3-5</th>
<th>6-8</th>
<th>9-10</th>
<th>11-12</th>
<th>13-15</th>
</tr>
</thead>
</table>
| Explain contact role at first visit. Review telephone contact numbers. Explain team concept. Introduce when needed – Child Life Care, Social Services, PM&R, and Dietary. Check immunization and flu vaccine status. Introduce MDA at second visit. | Annual visit
**Tier 1:**
Immunization review: pertussis, influenza, pneumococcal
Pulmonology health – PFT trend FVC
Canadian Occupational Performance measure (COPM)
Patient Activation Measure (PAM) – parent version
Kidscreen – starting age 8
**Tier 2:** # hospitalization in past 12 months with respiratory illness
**Tier 3:** # Diabetes screening/diagnosis
Presence of spinal deformity and contractures
BMI <18 or >29 |   |   |   |   | Continue measures |

<table>
<thead>
<tr>
<th>Ages 16-18</th>
<th>19-21</th>
<th>22-25</th>
<th>26-30</th>
<th>31-35</th>
<th>&gt;35</th>
</tr>
</thead>
</table>
| Transition to Lifetime care. Introduce new staff members and telephone contacts. Continue measures. | Bi-Annual visit with annual screening
Same measures as above with following modifications:
**Tier 1:**
Patient Activation Measure (PAM) – patient version
WHOQOL-BREF – starting with care at Lifetime
**Tier 3:** Depression screening (PHQ2 or 9 annually) |   |   |   |   |   |
Capturing Patient-Reported Psychosocial Outcomes: What Our Patient’s Tell Us

- Patient-Reported Outcome Measures (PROM) guide informed decision-making
- Know what is important to the patient
- Psychosocial measures are not consistently collected (Bushby et al, 2010)
- Patients face physical, cognitive, and emotionally challenges
- Experience with chronic pain, depression, and anxiety
- Barriers and disparities often limit interventions Knowing what is important to the patient is critical to optimum health.
- Organizations successfully implementing PROMs incorporate the patient voice into clinical decision-making.
Methodology & Study Design

• Quantitative design
• Convenience sample – 60 participants
  • Inclusion:
    – Confirmed neuromuscular diagnosis
    – Age 18 and older
  • Exclusion:
    – Decline research participation
• Pilot study – Feasibility to collect PROM data
• IRB approval

Marben, 2015
WHO Quality of Life-BREF

- World Health Organization project initiated in 1991
- Aim: develop an international cross-culturally comparable quality of life assessment instrument
- Assesses individual perceptions
  - Physical health
  - Psychological health
  - Social relationships
  - Environment
- 26 item self-administered questionnaire; 1-5 scale
- Cross-culturally valid
  (Skevington et al., 2004)

Sample Questions:
1. How would you rate your quality of life?
2. How satisfied are you with your health?
3. To what extent do you feel that physical pain prevents you from doing what you need to do?
4. How much do you need any medical treatment to function in your daily life?
5. How much do you enjoy life?
6. To what extent do you feel your life to be meaningful?
7. How well are you about to concentrate?
8. How safe do you feel in your daily life?
9. How healthy is your physical environment?
10. Do you have enough energy for everyday life?
11. Are you able to accept your bodily appearance?
12. How satisfied are you with your sleep?
13. How satisfied are you with yourself?
14. How satisfied are you with support from friends?
Patient Activation Measure

- Developed by University of Oregon – Judith Hibbard (Hibbard et al., 2005)
- Assesses 3 key domains & ability to self-manage
  - Knowledge
  - Skills
  - Confidence
- 13 item questionnaire
- Validated worldwide with 125 published studies
- Categorized into 4 levels activation
  1. Disengaged and overwhelmed (12-18%)
  2. Becoming aware, but still struggling (20-25%)
  3. Taking action (25-30%)
  4. Maintaining behaviors and pushing further (25-28%)
(Medicare patients in lower 2 levels 45-50%)

<table>
<thead>
<tr>
<th>Item</th>
<th>Disagree</th>
<th>Strongly Disagree</th>
<th>Agree</th>
<th>Strongly Agree</th>
<th>N/A</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. When all is said and done, I am the person who is responsible for taking care of my health</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>2. Taking an active role in my own health care is the most important thing that affects my health</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>3. I am confident I can help prevent or reduce problems associated with my health</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>4. I know what each of my prescribed medications do</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>5. I am confident that I can tell whether I need to go to the doctor or whether I can take care of a health problem myself</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>6. I am confident that I can tell a doctor concerns I have even when he or she does not ask</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>7. I am confident that I can follow through on medical treatments I may need to do at home</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>8. I understand my health problems and what causes them</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>9. I know what treatments are available for my health problems</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>10. I have been able to maintain (keep up with) lifestyle changes, like eating right or exercising</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>11. I know how to prevent problems with my health</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>12. I am confident I can figure out solutions when new problems arise with my health</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>13. I am confident that I can maintain lifestyle changes, like eating right and exercising, even during times of stress</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
</tbody>
</table>
Personal Health Questionnaire

- Depression risk screen – PHQ2+ and PHQ9
- Why screen for Depression?
  - Depressive symptoms can be fairly easily identified
  - Depression can be effectively treated
  - Untreated depression can exacerbate other medical conditions
  - To determine if remission from depressive symptoms occurs and is maintained
- Standardized and valid screening tool
- Scores 10 or greater (Q9- concern about self-harm) indicate risk; assessment with follow-up plan developed
- MNCM is a collaborative effort with the purpose of providing valid and reliable protocols for providers to use to improve patient care and outcomes.

Patient Health Questionnaire (PHQ-9)

Over the last 2 weeks have you been bothered by any of the following problems?

<table>
<thead>
<tr>
<th>Problem Description</th>
<th>Not at all (0)</th>
<th>Several days (1)</th>
<th>More than half the days (2)</th>
<th>Nearly every day (3)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Little interest or pleasure in doing things</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. Feeling down, depressed or hopeless</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. Trouble falling or staying asleep, or sleeping too much</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. Feeling tired or having little energy</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. Poor appetite or overeating</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. Feeling bad about yourself or that you are a failure or have let yourself or your family down</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7. Trouble concentrating on things, such as reading the newspaper or watching television</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8. Moving or speaking so slowly that other people could have noticed, or the opposite— being so fidgety or restless that you have been moving around a lot more than usual</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9. Thoughts that you would be better off dead, or of hurting yourself in some way</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

(Scoring for use by staff only) $\text{_____* _____* _____+ _____+}$

$=\text{Total Score}$ $\text{_______}$

NOTE: If you checked off any problems, how difficult have these problems made it for you to do your work, take care of things at home, or get along with other people?

Not difficult at all ______ Somewhat difficult ______ Very difficult ______ Extremely difficult ______

PHQ-9 is adapted from PRIME-MD TODAY, developed by Drs. Robert L. Spitzer, Janet W. Williams, Karl Klerke, and colleagues, with an educational grant from Pfizer Inc. For research information, contact Dr. Spitzer at rls@nysu.edu. PHQ9 Copyright © 1999 Pfizer Inc. All rights reserved. Reproduced with permission. PHQ9-A95 and PHQ9-ATM are trademarks of Pfizer Inc.
Feasibility: Time, Ease & Location

- **Time to Complete**
  - 10.65 min average
  - 3 – 30 min range
  - Most completed in 5-15 min (5.4 SD)

- **Location Completed**
  - 14 (23%) Waiting room
  - 46 (77%) Exam room

**Difficulty Rating (1 = easy)**

- 1 (37%)
- 2 (20%)
- 3 (18%)
- 4 (12%)
- 5 (7%)
- 6 (3%)
- 7-10 (3%)

75% subjects rate surveys as easy to complete
Aim: international recognized cross-culturally comparable quality of life assessment

26 item self-administered questionnaire

Rating scale 1-5 (not at all/very dissatisfied/very poor) to 5 (always/very satisfied/very good)

High score QOL 100 (highest satisfaction ≥75%)

WHOQOL-BREF Score (max score 100)

Mean 82

Frequency of Response

QOL score 82
Range 61-98
Psycho-social Outcome Measures

Patient Activation Measure (PAM)

**Aim:** Assess knowledge, skills, confidence & ability to self-manage (Hibbard et al., 2005)

13 item questionnaire; 4-scale options plus N/A

Categorized into 4 levels of activation

**Level 1** - Disengaged and overwhelmed (12-18%)

**Level 2** - Becoming aware, but still struggling (20-25%)

**Level 3** - Taking action (25-30%)

**Level 4** - Maintaining behaviors and pushing further (25-28%)

Patient Activation Measure by Condition

Average level 2.8 – becoming aware and taking action
Psycho-social Outcome Measures

Personal Health Questionnaire (PHQ 2 or 9)

Aim: screen for depression risk

- PHQ-2: two questions plus risk for self-harm
  - Responses: yes or no
  - If ‘yes’ to any questions administer PHQ-9

- PHQ-9: nine item questions (score 0-3) over past 2 weeks
  - If total score >9 then depression intervention plan
  - Screen for remission in 6 and 12 months

<table>
<thead>
<tr>
<th>Depression Risk</th>
<th>PHQ-2</th>
<th>PHQ-9</th>
</tr>
</thead>
<tbody>
<tr>
<td>Subjects</td>
<td>46 (76.7%)</td>
<td>14* (23.3%)</td>
</tr>
</tbody>
</table>

*Reporting to MN Community Measures
Organizations successfully implementing PROMs incorporate the patient voice into clinical decision-making.
Eating Assessment Tool-10 (EAT-10)

- Swallowing screening
- Early detection
- Validated patient self-report scale
- Threshold >3
- Speech evaluation referral
- Video swallow study
- Treatment plan

(Plowman et al., 2016)
Respiratory Control Plan

- Empower patients/caregivers to manage changes in pulmonary status
- Navigate within the Zones
  - Green: Routine treatments
  - Yellow: change in pulmonary status
  - Red: Pulmonary illness
- Decrease ER & hospitalizations

### Green Zone: I feel good “OK”

<table>
<thead>
<tr>
<th>Controller</th>
<th>Amount</th>
<th>How Often</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmicort</td>
<td>0.25mg</td>
<td>once daily</td>
</tr>
<tr>
<td></td>
<td>0.5mg</td>
<td>twice daily</td>
</tr>
<tr>
<td></td>
<td>1mg</td>
<td>twice daily</td>
</tr>
<tr>
<td>RESCUE</td>
<td>Vial</td>
<td>once daily</td>
</tr>
<tr>
<td></td>
<td>Puffs</td>
<td>Twice daily</td>
</tr>
</tbody>
</table>

**Airway clearance treatments**

Vest once Times a day
Lung Volume Recruitment Twice Times a day
Cough-Assist Four Times a day
Intrapulmonary Percussive Ventilator (IPV) As Needed

**Other Medications:**

**Yellow Zone: I DO NOT feel good “CAUTION”**

<table>
<thead>
<tr>
<th>Medicine</th>
<th>Amount</th>
<th>How Often</th>
</tr>
</thead>
<tbody>
<tr>
<td>Albuterol</td>
<td>Vials</td>
<td>Times per day and Every _ hours as needed</td>
</tr>
<tr>
<td>Xopenex</td>
<td>Puffs</td>
<td>Times per day</td>
</tr>
<tr>
<td>Pulmicort</td>
<td>0.25mg</td>
<td>Times per day</td>
</tr>
<tr>
<td></td>
<td>0.5mg</td>
<td>Times per day</td>
</tr>
<tr>
<td></td>
<td>1mg</td>
<td>Times per day</td>
</tr>
</tbody>
</table>

**Airway clearance treatments**

Vest once Times a day
Lung Volume Recruitment Twice Times a day
Cough-Assist Four Times a day
IPV As Needed

**Other Medications:**

**Red Zone: I feel awful “DANGER”**

<table>
<thead>
<tr>
<th>Medicine</th>
<th>Amount</th>
<th>How Often</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prednisone</td>
<td></td>
<td>2 times daily for ___ days</td>
</tr>
<tr>
<td>Prednisolone 15mg/5mL</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Airway clearance treatments**

Vest once Times a day
Lung Volume Recruitment Twice Times a day
Cough-Assist Four Times a day
IPV As Needed

**Other Medications:**

**FOR RAPID INCREASE IN SYMPTOMS MOVE TO RED ZONE MEDICINE**

- You are struggling to breathe
- There is simply no clear improvement
- You cannot walk or talk in full sentences
- Your lips or finger nails are blue

**Health Care Provider Signature**

**Date**

**Parent/Guardian Signature**

**Date**

**Return to Clinic in:** ___ days ___ weeks ___ months ___ years

**Parent/ Guardian Signature**

**Date**

**END DUCHENNE.ORG**
Conclusion

– With early diagnosis & coordinated plan of care individuals with muscular dystrophy can reach their full potential in education, employment, and independence.

– Interventions have led to improvements in function, quality of life, health, and longevity

(Bray, Bundy, Ryan, North, & Burns, 2010; Holloway et al., 2008; McDonald, 2013)
We have more questions, better questions, and a few suggestions and a few answers....

Contact: kmarben@gillettechildrens.com
References


