Transitions in Care in SMA

Case contributor and commentary:
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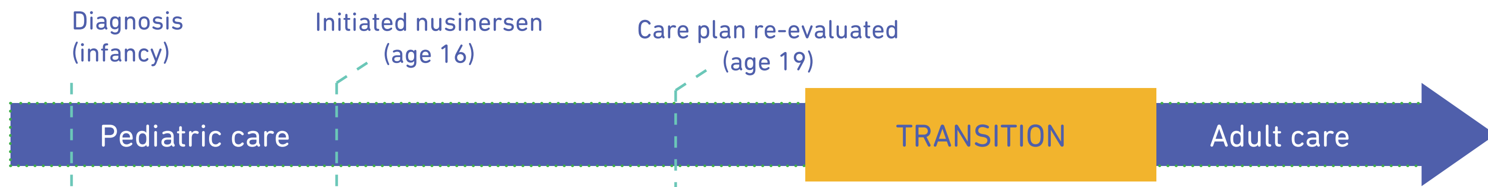
Patient #3: Supporting social/emotional health during transition in care

19-year-old female

Case Timeline



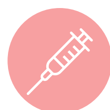
Early life

The patient received a diagnosis during her early life

- Clinical diagnosis of SMA type 2 was made at infancy, but *SMN* copy number was unknown
- Never achieved ambulation, but was able to sit, supporting a clinical diagnosis of SMA type 2

Age

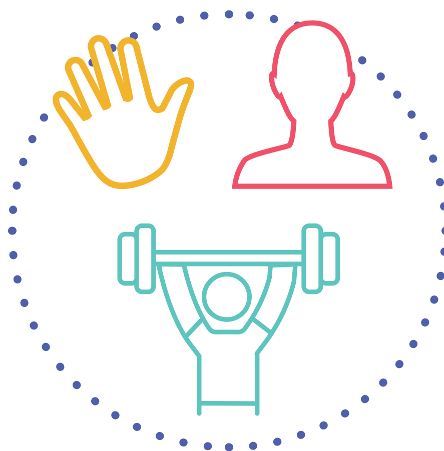
16 - 19



Initiation of disease-modifying treatment

The patient began nusinersen therapy at age 16 and tolerated it well

- Contemplated a switch to risdiplam because of the increased difficulty of frequent lumbar punctures
- Continued to show improved strength after starting nusinersen
  - Improved hand strength
  - Increased ability to lift objects a little easier/higher
  - Improved neck strength



MDA Muscular Dystrophy Association

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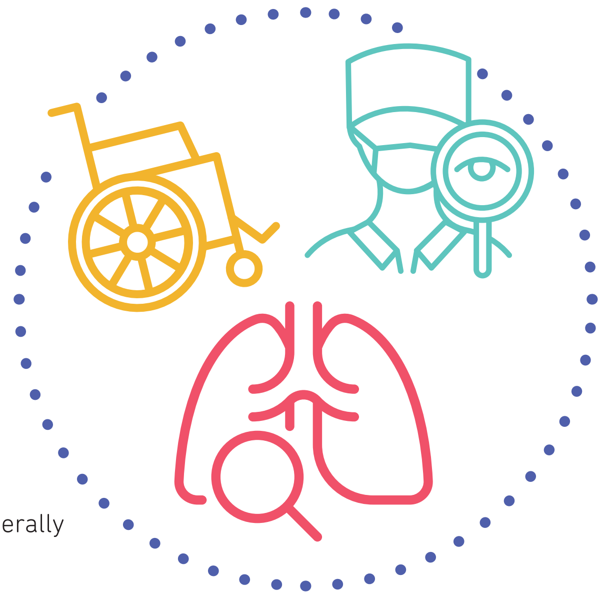
Review of health status



Nutrition

- Ate by mouth and lost some weight
- Had a gastrostomy tube used mostly for medications and fluids
- Followed with team dietitian

Respiratory status

- No respiratory concerns
- Slept with BiPAP at night and for naps
- Used Cough Assist intermittently
- Regularly visited with a pulmonologist

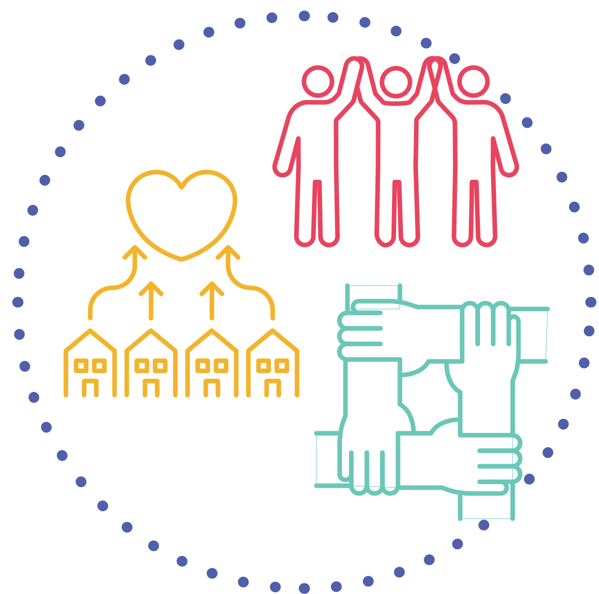
Musculoskeletal status

- In a motorized wheelchair full-time
- Intermittently received PT
- Had upper and lower extremity contractures and pain in her hips, bilaterally

General health

- Good skin integrity

Review of social and lifestyle status



Living situation

- Lived at home with mother and younger unaffected brother
- Had aides for 16 hours/day that helped with activities of daily living and accompanied her to appointments

Social health

- Very active/busy lifestyle
- Had a boyfriend that lived in California
  - Met online and talked every day
  - Were planning a visit soon
  - Patient was interested in family planning options

Mental health

- History of intermittent depression, but seemed to be improving

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Age
19

Based on diagnosis of type 2 SMA, the following multidisciplinary evaluation and management plan was implemented.

	Evaluation	Symptom management	Treatment goals
Physical therapy (PT)	» CHOP INTEND score and Hammersmith score were regressing slowly over time	» Patient was instructed in stretching during visit and assured proper fit of current motorized wheelchair	» Encourage return to weekly outpatient PT » Encourage ongoing stretching and range of motion exercises at home, as much as possible » Recommend new leg braces
Occupational therapy (OT)	» Patient had increased difficulty raising hands to mouth to eat, drink, and brush teeth	» Demonstrated use of different devices (including a Wilmington robotic exoskeleton (WREX)) for easier use with arms » Discussed aqua therapy	» Schedule trial of WREX » Consider and identify resources for local aquatherapy
Nutrition	» Reviewed daily dietary intake	» Discussed ways to increase calories in diet (e.g. - adding cream to foods, adding in shakes, eating higher calorie foods, etc.) » Initiated conversation about tube feeds	» Increase weight by 2-3 lbs by the next visit » Schedule a barium swallow study to assess safety » Check Vitamin D level and follow up if supplementation is needed
Social Work	» Explored possibilities for meeting boyfriend and potential future plans	» Discussed plan for going to OB/GYN and options for birth control	» Schedule OB/Gyn appointment » Give resources to encourage healthy relationships/safe sex practices » Discuss importance of birth control while on risdiplam
Neurology	» Evaluated current progress while maintained on nusinersen	» Discussed option for switching to risdiplam, including potential for future pregnancy/family planning	» Start the process for switching to risdiplam as daily medication is desired without the need for LPs » Reassess strength and progress in upcoming months » Monitor labs when starting risdiplam » Schedule eye exam
Pulmonary	» Mild changes in pulmonary function testing (PFTs)	» Reviewed current BiPAP settings	» Change BiPAP settings slightly » Increase use of Cough Assist to twice daily for pulmonary clearance
Orthopedics	» Had spinal fusion surgery at approximately 8 years old	» Due for visit with orthopedics to address hip pain	» Reduce hip pain without the need for surgical intervention
Cardiology	» Had palpitations in the past » Cardiac work-up was normal	» Told to return if symptoms reappear or as needed	» Return if symptoms reappear/as needed

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Transition to adult care

During the transition from a pediatric clinic to an adult care clinic, a social worker reviewed aspects of adult care and assessed the patient's competence and readiness for transition using the "Readiness Scale."

Assessment of Patient Readiness and Competency in Medical Decision-Making			
Medical Providers/Care » Is familiar with all providers and knows how to contact them	**Medications** » Has current list of medications updated regularly by aides	**Sexuality** » Interested in initiating a sexual relationship » Needs to explore options for birth control and family planning	**Mental Health** » Sees a therapist » Is on medication for depression
Transportation » Knows how to arrange for transportation	**Emergencies** » Feels prepared about whom to call in an emergency » Local police and fire station have her information on file	**Insurance** » Has insurance information on file » Is unsure if she needs to change anything in her transition to adult care, but asks appropriate questions	**Financial aspects** » Manages her own finances with assistance from her mother

A medical care transition plan was documented and communicated to the adult care team.

- Paperwork was reviewed and signed by patient/guardian to allow release of information to the adult care team
- Social worker prepared transition packet and communicated with the adult care team to schedule first appointment at the adult clinic
- Plan was relayed back to the patient at the next clinic visit
- Adult care team contacted the patient to schedule the first appointment

The nurse practitioner met with the patient to discuss goals for care and advance care planning wishes using the *Voicing My Choices* conversation tool.[1]

Patient responses: *Voicing My Choices*

How I want to be comforted
- Patient wanted favorite blanket if ever in the hospital
- Could not be without her phone and iPad
- Loved to listen to music and paint

How I would like to be supported
- Wanted family and friends around her, especially if she was feeling sick
- Wanted to get married and have her husband with her at all times

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Patient responses: *Voicing My Choices* (continued)

Who I want to make my medical care decisions if I cannot make them on my own

- Current: mother, younger brother second to mother
- Future: husband

The types of life support treatment I want or do not want

- Would NOT want CPR if her heart were to stop
- May want increased respiratory support, depending on the circumstances
- Wanted to receive artificial nutrition and hydration and blood transfusions, if needed

What I would like my family and friends to know about me

- Patient stated:
  - "I love them and am grateful to them for always taking care of me and treating me as an equal."
  - "I forgive my father even though I have never met him."

My spiritual thoughts and wishes

- Religion was not too important to the patient, but she believes in a higher power and heaven
- She would want people to pray for her if she was sick

The advance care plan was documented by the nurse practitioner.

- *Voicing My Choices* was scanned into the chart and the original document mailed to the patient
- The patient wished to have Advance Directive/Do Not Attempt Resuscitate (DNAR) orders in place
- Was completed and placed in the chart and an out-of-hospital DNAR was completed for home

Key learning points

- Physical, mental, emotional, and social aspects of care are important
- Interdisciplinary team members all play an important part in contributing to the well-being of each patient and helping to meet his/her individual needs
- Adolescents and young adults should be a part of their care as they transition to adult care and their readiness should be assessed and a comprehensive plan developed to meet all domains of care
- Discussion goals of care/advance care planning is an important aspect of care for adolescents and young adults

References

1. Zadeh S, Pao M, Wiener L. Opening end-of-life discussions: How to introduce Voicing My CHOiCESTM, an advance care planning guide for adolescents and young adults. Palliative and Supportive Care. 2015;13(3):591-599. doi:10.1017/S1478951514000054