Chapter 8
Transition Care for Adolescents and Young Adults with Neuromuscular Disease and Chronic Pulmonary Care Needs

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Case JW is a 19-year-old woman with a history of spinal muscular atrophy (SMA) type II. Her symptoms started at 12 months old, when she had difficulty in sitting without support. She has scoliosis and chest wall deformities requiring spinal fusion. She was able to crawl but never was able to walk unaided. She uses a motorized wheelchair for mobility. She was treated as a child at a tertiary children's hospital. She was found to have respiratory muscle weakness with a forced vital capacity (FVC) of 0.51 L, 26% predicted, and has been using nocturnal bi-level positive pressure ventilation for support. She uses a mechanical insufflator/exsufflator device for cough assistance. She will now be attending college and she presents to the adult pulmonary clinic to establish care and long-term follow-up. She has to deal with changing insurance and moving to a new state, as well as getting to know a new doctor and clinic.

Introduction

Chronic neuromuscular respiratory failure is a consequence of a number of conditions that first present in childhood, such as myotonic dystrophy and spinal muscular atrophy (SMA). Many of these patients are treated at pediatric hospital specialty clinics, such as Shriners Hospitals or Muscular Dystrophy Association (MDA) clinics, although some patients may be followed by individual pediatric providers

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(subspecialty and primary care). Multidisciplinary clinics have provided these children comprehensive care including social work, dieticians, physiatrists, neurologists, pulmonologists, and others. This care has extended the life span for these conditions and many patients are now living into adulthood [8]. Though epidemiological data is quite limited for most neuromuscular diseases, incidence of SMA is cited at 1 per 6,000–10,000 live births with prevalence of 1–2 per 100,000 persons. Duchenne and Becker muscular dystrophies come to an incidence of 1 per 3500–5000 male births, meaning approximately 400–600 affected males born in the USA annually [12, 13, 15, 21, 24]. The frequencies of many conditions, including rare diseases, are often unknown as there are no active registries.

Advancements in mechanical ventilators and mechanical insufflation/exufflation devices over the last 30 plus years have made them smaller and easier to use. These advances have facilitated the transfer of care for children with chronic neuromuscular respiratory failure to the home environment instead of long-term residency in medical institutions [27]. Improvements in mask technology and mouthpiece ventilation have allowed patients to avoid tracheostomy as well [3]. Patients with Duchenne muscular dystrophy (DMD) and SMA are now living longer. There is drastic variation on life expectancy across the spectrum of clinical manifestations in these conditions. Over the past couple of decades, patients with DMD experienced an increase in life expectancy of more than 10 years, with now routinely living into third decade of life [25]. People with SMA are experiencing an unprecedented and undefined expansion of life expectancy with new disease-modifying agents. With this increased survival, the need for organized transition from pediatric to adult providers has become more important [2, 7, 16, 31].

There are a number of transitions in the process of going from pediatric to adult pulmonary care for these patients who may be device-dependent. The structure of pulmonary training in the USA also plays a role in the transition as well. Providers are typically trained in pediatrics or internal medicine first before subspecialization, meaning that there are not classically pulmonologists cross-trained in adult and pediatric medicine. This requires patients to transition from one practice environment to the other with respect to respiratory management. Exact timing has been debated for this transfer of care. Challenges include having adult providers that are familiar with the underlying neuromuscular conditions and willing to engage with these complex patients. Pediatric tertiary clinics also typically have more support staff and services, such as social work, that are often lacking in standard adult pulmonary clinics. Replacing these services on the adult side of the transition is challenging and can leave patients and caregivers without as many resources.

Coordinating a planned transition has been proposed by a number of organizations. The Got Transition website is a collaborative project designed to help guide the process for both pediatric and adult providers. It has many helpful resources, but is not specific for subspecialties. In this chapter, we will discuss patient-centered models that are specific to pediatric to adult transition for patients who are ventilator-dependent due to chronic neuromuscular respiratory failure.

Logistics

As JW is heading off to college, she certainly has medical factors to consider, but there are also several logistical issues that will impact her next steps. She needs to find providers to help adjust her respiratory support and medications, but she also needs to find housing that can accommodate her devices and figure out transportation on campus. The logistical challenges faced by adolescents and young adults (AYA) transitioning with chronic disease, such as MD, are as important, arguably more so, than the medical aspects of their transition and require attention and support through the process. Patients' quality of life and reaching maximal functional independence can benefit greatly from involvement of a multidisciplinary support team familiar with community resources and the issues common in this population.

Ideally, for these patients, social workers (SW) or case managers familiar with local resources and policy are an integral part of clinical care. If this is not feasible for a clinic, these providers should be made as available as possible, and regularly updated lists of community organizations should be easily accessible to patients and families. Care managers and social workers who are well informed of community resources for people with neuromuscular disorders (NMDs) can provide invaluable insight and assistance in these situations and ideally should be a central part of the multidisciplinary teams caring for children and adults.

One of the major transitions that people with NMD experience is moving to independent housing or, alternatively, navigating the transition into a young adult in the family home. This change may be prompted by starting higher education, entering the workforce or a vocational program, or aging out of the public school system. For those with significant cognitive delay or severe physical impairment, living independently may not be a realistic option [4]. Others are best served by transitioning to a group home or transitional/supportive environment separate from the family home. Many will be able to transition to independent housing, which comes with its own unique challenges and rewards. It is extremely important to ascertain the wishes, fears, and goals of patients and families when addressing changes to the living environment [18]. Many patients and families find attaining physical independence to be a vital aspect of the adult experience, and they place great value in facing any challenges necessary to allow for independent living. Others may not find this to be their most important goal and would prefer to focus their energy on other areas of achievement while remaining in the family home. Providers should aim to focus encouragement, support, and resources on what is most important to the patient, which may not necessarily fit with traditional models defining what it means to be a high-functioning adult [30].

Aging into young adulthood in the same living environment still requires adjustment by the patient and family with regard to roles in the home, independence and autonomy in engaging in social time and self-care, and social and romantic relationships as age appropriate. Even more relevant than objective age is the patient's cognitive and emotional maturity, which does not match chronological age for many patients with chronic illness. These young adults may also be potentially taking

more of a role in the financial and chore support of the household. Even if the patient's physical location is not independent, they can still progress in functional and social independence. This is another area in which adequate counseling and social work support can be of great value to patients and families. Many children grow up reliant on their caregivers for much more (and for much longer) than the average child; self-care issues such as toileting, bathing, medication regimens, self-cathing, wound care, and obtaining nutrition (whether orally or via enteral support) are complex aspects of care in a developing adolescent and warrant dedicated attention from providers to ensure that patients take on as much autonomy as safely possible. It is also worthy of time and care as some patients may need more, rather than less, assistance as they age and experience disease progression.

Patients and families not only need to navigate the in-home social changes during this time, but also the logistical support of a growing and maturing child in the home. Depending on the state of residence, there are varying levels of support available for young adults needing environmental modification, assistive devices, and support and recreational groups. The amount of public financial support for patients with NMD also varies by state [30]. Many AYA must navigate a profound change in resources as they age out of school-centered supports [23]. Under the Individuals with Disabilities Education Act of 1975, students must be provided with free, appropriate public education in the least restrictive environment possible. These services begin with early intervention at ages 0–2 years and special education services ages 3–21 years. Also under this law, students must have a personal transition plan by 16 years of age and work toward developing functional skills [33]. Though many students rely heavily on the educational environment and feel the loss of leaving high school, these transition plans and skills can be very useful in moving into the vocational training realm, as well as into various other levels of community engagement, education, and employment.

The path to independent living in the community and/or utilizing vocational training can be complex and confusing. Resources vary greatly between communities, requiring more support from social work and community organizations to identify housing options. On an individual basis, factors such as accommodations for degree of disability (elevators, ADA-friendly countertops and bathrooms, etc.) and adequate accommodations for devices (electrical plugs, physical space in bedrooms and bathrooms, etc.) need to be addressed. It is also important to find an environment appropriate for the person's level of social and psychological abilities and comfort. There are numerous community, federal, state, and local government programs that can facilitate services, but knowing the specifics of the patient's local area is key. Some potentially helpful programs include Assistive Technology Resource Centers, the local Department of Health and Human Services (which can be helpful in identifying local resources, completing registrations, and getting on wait lists), and local branches of disability services. Home caregivers can be very helpful, with payment through community groups, insurance, or private pay.

Vocational training is a particularly valuable opportunity for many people with chronic conditions, as many are excellent candidates for adapted training and work environments despite deficits in academic, social, and physical functioning [26]. If

joining the traditional workforce, patients should be encouraged to find employment that fulfills their interests and needs while balancing the needs of their medical conditions (transportation, physicality of the job, time needed for appointments and treatments, etc.). They should be aware of legal protections for people with disabilities and familiarize themselves with available recourse if being treated unfairly. The Rehabilitation Act of 1973 initiated the movement of equal rights and protections for people with disabilities and has been followed by several other legal statutes that support this population. Under this Act, organizations receiving federal money or that are federal programs cannot discriminate on the basis of disability [36]. The Americans with Disabilities Act of 1990 was a huge step forward for this population, mandating modifications to buildings and public spaces to make them accessible to all and supporting services for those with vision or hearing impairment [1]. The following resources may be a helpful place to start for questions and support:

- American Civil Liberties Union Disability Rights Program, www.aclu.org/disability
- Disability.gov
- https://www.ada.gov/cguide.htm

Transportation also poses unique challenges. If feasible and desired, providers can assist in facilitating acquiring a driver's license and vehicle. Again, working with social work and community resources to find best solutions for transportation needs is an important part of transition planning. Several forms of public transportation may suffice in areas with adequate routes and schedules. Smaller programs for individuals with specific needs may be accessible through community organizations. Much of the discussion about transportation goals (license, etc.) and solving problems, like how to get to school, work, and social activities, should be ongoing through the early years of the transition process but may need refinement as young adults gain more independence and autonomy.

People seeking traditional higher education may benefit from a more directed line of next steps involving student housing or other housing options based through their institution, with clearly designated staff to help students navigate institution-sponsored housing options. When entering higher education, any student (and especially those with any physical or educational disabilities) is required to take over the role of being their own advocate. While most settings in the pediatric world allow for a caregiver or provider to intervene on behalf of the patient, higher education requires a greater degree of autonomy and self-reliance. Students need to have a clear understanding of their role in this arena and acquire the self-help tools they need to reach out to school officials to register their disability and engage in sufficient support services. Many students make the mistake of trying to adapt on their own to the new environment and end up frustrated with a wasted semester before they engage with the school to obtain appropriate accommodations. For example, students with audio or visual disabilities may benefit from extra time for exams,

note writing services, or vision/hearing devices during lectures. Limitations with fine motor skills may require adaptations to use computers or completing written assignments or exams. Students dependent on a wheelchair may need assistance with building a schedule that coincides with wheelchair-accessible bus routes, ADA-accessible classrooms and elevators, and adequate time between classes to get across campus safely and complete any medication or treatment regimens. Similarly, student housing considerations will need to include ADA-accessible facilities and sufficient space and plug-ins, etc., for equipment. Below are some potentially helpful sources of information:

- FERPA, www2.ed.gov, regarding privacy of student records
- IDEA, https://sites.ed.gov/idea/

So how does JW begin to tackle these challenges to reach her goals? Thankfully, in her situation, she has benefitted from strong support from her family and clinic providers through her adolescence. She has not learned to drive and is not interested in pursuing that goal. She has been able to use public transportation in her home city but is concerned that the bus routes in her new college town may not fit her class schedule. She is also concerned that the usual freshman dorms do not have adequate elevator access or bathroom and dorm room adaptations that will allow her to live with her peers. She is also concerned about moving between classes fast enough and completing exams in the regularly allotted time. In high school, she had an Individualized Education Plan (IEP) that accommodated her educational experience to allow her the best chance of success. Without that, she is nervous about not succeeding in her first year. Luckily, her college has an office that actively supports students with disabilities. Using the self-help skills she developed during her transition process, she reaches out to the office well before the start of the semester. This office is able to help her secure student housing in a different building that meets her needs and has the space for a personal caregiver to help her with treatments and care a couple of times per week. She arranges a class schedule that has adequate time to support her mobility needs and intentional breaks to allow her to perform her medical treatments and take medications. She articulates what accommodations she needs from her professors ahead of time, with letters of support as needed from her medical providers. Finally, JW finds a local organization that can give her rides to/from clinic appointments and the grocery store. Her parents are able to pick her up for school breaks and the occasional weekend. Clearly, the process of starting college is much more complex for these patients. They likely have the same fears and worries as typical new students, with the addition of fears and worries related to their disease process. Extra support and preparation can go a long way in setting our patients up for success as they navigate the adult world.

Insurance coverage continues to be a critical factor in determining the location, providers, and the extent of resources available to all patients in the USA, with

particular importance to patients with chronic conditions and high levels of medical need. As patients enter adolescence, it is critical that providers incorporate regular and detailed discussions about future insurance coverage into appointments and transition planning. Patients have several potential sources of insurance. Under the Affordable Care Act (ACA), many patients can remain on their parents' insurance policy until age 26. There are potential state-by-state continuations of coverage if the dependent is unable to self-support. ACA legislation also eliminated exclusions for pre-existing conditions and caps on lifetime coverage. Also facilitated under the ACA, personal policies can be found on the Healthcare Exchange [44]. Many patients with a NMD qualify for benefits under Supplemental Security Income (SSI) and Social Security Disability Insurance (SSDI), which correlates with qualifying for Medicaid coverage. In many states, qualifying for SSI automatically qualifies for Medicaid. In 11 states, an additional application is required. Patients can potentially get Medicaid in states that expanded under ACA if they do not qualify for SSI. Patients enrolled in higher education may have access to private insurance through their institution. Discussions about plans for insurance coverage as patients age into adulthood should begin early in the transition process and involve patients, parents, and the care team with the goal of avoiding any lapses in care. As patients are transitioning to their adult care teams, attention should be given to availability of appropriate in-network providers wherever patients will be living (college, home, etc.).

Most people with a NMD meet criteria for SSI and SSDI based on their diagnosis. Hopefully, patients have been enrolled as children and have been receiving these benefits since close to the time of their diagnosis. If not, families should be directed to resources to help them register. Qualification for SSI varies by state, but in general, one can be eligible with a diagnosis that significantly limits function and/or if a person meets certain income requirements. As patients approach their 18th birthday, they will need to be prepared to undergo a benefit review to see if they remain eligible for SSI. The patient's childhood condition may fall under Medical Listing/Blue Book and automatically denote qualification, or the review may need to demonstrate severity to meet listing to qualify. For this review, SSI officials usually need medical records for the prior year, and medical providers can also offer a letter of support in the application [10]. In most states, if an individual qualifies for SSI, they also qualify for Medicaid insurance coverage. Many of these patients may also qualify for SSDI benefits, but these are limited to citizens who have a work history. Again, patients need to meet certain medical requirements [9]. SSDI coverage is particularly helpful for patients who are able to work part of their adult lives but lose the ability to maintain gainful employment due to progression of disease. These patients should be directed to start the SSDI process as soon as unable to work, as a waiting period exists for benefits to start. After 24 months of receiving benefits and the waiting period (total of 29 months), patients are also eligible to start receiving Medicare benefits. A few conditions automatically qualify for Medicare, with ALS being most relevant to this population [42]. There are certain supports as well as restrictions for patients receiving SSI and SSDI benefits with regard to the level of gainful employment in which they can engage while still receiving benefits, and

patients to whom this applies should be familiar with these earning caps. Below are some potentially helpful links:

- www.ssa.gov
- https://www.ssa.gov/benefits/disability/
- https://www.ssa.gov/disability/professionals/bluebook/general-info.htm
- ACA US Department of Health and Human Services, https://www.healthcare.gov/, https://www.hhs.gov/healthcare/about-the-aca/index.html
- https://www.hhs.gov/answers/medicare-and-medicaid/who-is-elibible-for-medicare/index.html

Many of the conditions causing neuromuscular disability also entail neuropsychiatric conditions, some involving changes to cognitive function [4], such as frontotemporal dementia/executive function changes in amyotrophic lateral sclerosis (ALS) and potential cognitive involvement in cerebral palsy (CP) [19]. Others have strong associations, such as autism spectrum disorder (ASD) and attention-deficit/hyperactivity disorder (ADHD), in DMD [29]. Many patients will also experience secondary effects of their condition. In the setting of medical complexity and dependence on multiple devices and treatments, frequent hospitalizations, and experiencing childhood as a chronically ill person, many adolescents and young adults have delayed psychosocial development and maturity [14, 26]. An important role of transition planning is to educate patients and families about steps toward independence and enforce practices that enable patients to take over their own care (talk directly to patients, interview independently, graded ownership of care and medical information). For some, dedicated efforts to facilitate independence are able to help them attain a desired level of independence. For others, "full independence" in the traditional sense is not a realistic goal, and an optimistic but realistic approach to their goals is needed [4]. It is very important to discuss with patients and parents what their goal adulthood looks like. Some patients will desire as "normal" of a life as possible, while others will be very content with graded independence. The traditional view of achieving independent adulthood may or may not apply, and providers need to keep an open mind in discussing what matters most to patients, then focusing on these goals as better markers of optimized outcomes.

Many patients/families will also need to address guardianship, which primarily refers to appointing a person(s) to make medical decisions for someone (sometimes employed along with conservatorship, which primarily refers to a legally appointed person responsible for the financial decisions of an impaired individual). The need for guardianship should be assessed early on in adolescence, as the process requires time, and can be much more challenging after the patient has reached adulthood. Guardianship is a parallel concept to competency, both being legal designations of whether or not an individual can make their own decisions. Capacity, on the other hand, can be fluid and case-by-case with different decisions/situations. This can be assessed by clinical providers and can change day to day. To have capacity, the

patient needs to be able to understand the risk/benefit/alternative of treatment options and reason with this information to make and communicate their decision. It is important to establish wishes, roles, and documentation before an emergency arises that limits capacity. Shared decision-making should be used whenever possible if independent decision-making is not feasible (*disability*). Legal support varies by state, but assistance can be found through Supported Decision-Making, ACLU Disability Rights Program, and National Resource Center for Supported Decision-Making, or the Quality Trust for Individuals with Disabilities. If guardianship is warranted, this process is completed through a petition to the court, and the specifics vary by state. Through the National Guardianship Association, one can look for state organizations which can help navigate the process, as well as when and how to start. Whenever possible, patient preferences, input, and autonomy should be respected, and patient autonomy should be preserved as much as possible. Below are resources that may be helpful:

- supporteddecisionmaking.org – National Resource Center for Supported Decision-Making
- dcqualitytrust.org – Quality Trust for Individuals with Disabilities
- www.guardianship.org – National Guardianship Association

While some patients may be able to reach full independence, or nearly so, most will still have some dependence on caregivers. An additional consideration as patients age through adolescence and young adulthood is that their caregivers, often parents, are also aging and facing their own medical and functional changes. Caregiver responsibilities may be taken over by paid caregivers, other family members or friends, or a significant other of the patient. This change can be challenging emotionally, logistically, and financially for both patients and caregivers. Providers can help patients and families navigate these situations again by relying on multidisciplinary support from social workers [30] and counselors/mental health providers and by giving dedicated attention to these topics in transition planning visits. Parents and primary caregivers should be counseled to consider their child's long-term care in estate planning and should arrange for caregivers/support in case of unexpected death or debility.

Social/Emotional/Functional Aspects

As part of normal development, patients progress through stages to become independent from their parents and caregivers. This complex process may be delayed when these patients are dependent for many of their activities of daily living and may be further complicated when the patients have cognitive deficits [4]. In *Living with MD1*, the mother of four patients with myotonic dystrophy 1 details the

struggles that her sons and daughter went through during their adolescence and those that persist into adulthood [43]. Patients with this condition (or other NMD) may desire to have normal friendships and romantic relationships despite being dependent for activities of daily living and needing to use positive pressure ventilation at night. Relationships can be particularly challenging when the patient has a disease that will be life-limiting [18, 41]. Some patients have joined condition-specific support groups online that help patients navigate daily life. Support groups can be helpful for caregivers as they navigate the complex medical world. Social workers can be helpful for providing further local and national resources. This aspect of growth and development in AYA with NMD highlights the importance of supportive medical and social environments, as well as an area of life that contributes greatly to quality of life and warrants adequate mental health and medical support.

It can be challenging for providers to help patients become more independent both socially and medically. Got Transition recommends that when the patient reaches age 12, joint discussions should begin with patients and caregivers to plan these transitions. Taking responsibility for their own medical care is an important step for some patients, while others may not have the cognitive or physical capability to take complete responsibility for their medical care.

The subject of our case study has significant mobility issues and requires caregiver assistance with some activities of daily living such as wheelchair transfers. She noted that her parents taught her to be a vocal advocate for her care even though she was not able to do everything herself. She had hired caregivers and trained them with proper protocols. She reports that doing this at a young age gave her confidence and control over her condition. Importantly, she was able to standardize her care between caregivers. One of her challenges with transitioning to college was transportation. She uses a motorized wheelchair and the bus transportation system, but some portions of roads and sidewalks were difficult to navigate. She also had difficulty with new buildings that could not accommodate her wheelchair. This required considerable advanced planning to adapt to the university environment. Furthermore, upgrading and adjusting her wheelchair as needed has been a challenge with insurance coverage.

Her transition also entailed the challenge of hiring new qualified caregivers. She was able to hire students from a local nursing school, and they in return gained experience and were compensated for their services. She is a very bright student and did well academically in high school. She was interested in journalism study for college. Prior to selecting her university, she inquired about accommodations for both physical and academic options. She needed more time for transportation around campus because some buses did not have wheelchair capability. Location of classes was also important because she would need to find elevator access and adequate room for her wheelchair. She also felt mobility and access was key for being able to socialize with other students and to feel like she was a part of the community. She wanted to be as independent as possible and succeed like other students. Sometimes these challenges would be difficult and our patient would struggle emotionally. She contacted the clinic social worker for support and was able to receive

counseling, although the first recommendation she received was for a counselor in a building that was not wheelchair accessible.

Successful transition for a number of patients requires using resources such as counseling, social work, and in some cases palliative care. Pediatric palliative care has had different models, depending on location and institution. Palliative care on the adult side is often used when patients are within 6 months of death or making difficult care decisions. It has been observed that pediatric palliative care patients are followed more long-term and for more symptom management. Early and consistent involvement of palliative care (via primary providers or a consulting specialty team) can help to address psychosocial distress as well as physical symptoms of disease and treatment. Early involvement also is known to enhance quality of life and minimize aggressive end of life interventions when not consistent with patient goals [35]. Additionally, these patients benefit from extra support in developing coping skills in the face of progressive illness [41]. Palliative care providers receive extra training to help manage symptoms and support quality of life while adequately supporting the patient's developing psychosocial abilities and needs.

In the multidisciplinary clinics at the University of Michigan and at the University of Utah, we have used a combination of social workers, psychiatrists, therapists, and palliative care to cover the spectrum of needs. On average about 16 patients per year transitioned from pediatric providers to the adult clinic (personal communication from the author). Advanced planning is needed for major life decisions and for facing the reality of aging caregivers. Institutional support is key in providing resources. Full-time equivalents can be shared between different clinics to help support the cost of these providers and support staff. The resources skilled support staff (such as dieticians, care managers, PT/OT, etc.) provide for these patients and providers are invaluable.

Medical Transition

Ideally, the care environment for adults with neuromuscular disease would mirror that of pediatric multidisciplinary clinics. Though data in this area is sparse, patients with NMD do tend to have better outcomes and higher levels of societal function if involved in a multidisciplinary clinical environment [5, 38, 41]. If that is not possible, close communication between the pediatric and adult providers including social work and palliative care is key. Changes in equipment may also happen with pediatric to adult transition, such as tracheostomy placement, for patients that use invasive ventilation. An otolaryngology provider could follow them long-term or they may need a new provider. New insurance coverage and new DME companies will often be part of the transition as well. One example we have noted is that vest therapy, as a mucus clearance device, is favored more frequently for pediatric patients than adult patients.

As these patients transition, there also needs to be planning discussions about general health and function, including reproductive health. A genetic counselor

could be helpful for this as well. Mental health support is extremely important and also an inadequately addressed area of wellbeing for these patients [20, 41]. Aside from multiple issues specific to the NMD involved [6], AYA patients still need the normal screening and counseling appropriate for their age group, which can usually be managed by an involved primary care provider within the multidisciplinary medical home or in the community [34, 45].

Transition Stages/Process

Got Transition is a national program focused on healthcare transition with many helpful resources that are easily accessible online from the homepage of gottransition.org. It is a cooperative agreement between the National Alliance to Advance Adolescent Health and the Maternal and Child Health Bureau. The content of Got Transition is aimed to serve healthcare providers, patients, and families. Within the website are several documents to support providers in making a transition policy, tracking individual progress during the process as well as assessing readiness and planning for transition, carrying out the actual transition to adult providers, welcome/orientation visits, and tracking outcomes. There are distinct sets of resources for pediatric providers, accepting adult providers, and those who may be taking care of the patient across the life span (i.e., family medicine or internal medicine-pediatrics-trained physicians). Got Transition also has information about provider education and many patient topics such as guardianship, insurance, education/employment, and resources for patients. This is an excellent resource for providers, patients, and families and contains a well-curated body of information for reference and program development with much of the content also available in Spanish [17].

Starting around age 12, patients should enter into the transition process. Initial discussions can involve introduction of a transition policy, which generally includes when and how this process happens in clinic, as well as expectations and goals for providers, parents, and patients. During the transition process, patients then perform repeated self-assessments to assess readiness and progress. Over time, patients should proceed stepwise through a checklist adapted to the clinic and to the patient's specific needs and disease process. It is expected that this process will take several years, and variability between patients is also to be expected. Written (or electronic) self-assessments during clinic visits can allow patients and providers to monitor objective changes over time as patients progress in their readiness. It is time to formulate a plan for the actual transition to adult care once a patient (1) has completed the checklist or is nearing completion, (2) has a good understanding of their medical situation and needs, and (3) is progressing in self-care and self-advocacy skills (monitored by self-assessments). The patient then needs to gather a medical summary, including plans of care centered on patient goals, prepare an emergency plan, identify adult providers, and schedule first visits with those providers. Such a plan will require substantial individualized adaptation for patients with intellectual disability or severe functional impairments. Providers and parents should be discussing

more global issues such as guardianship/conservatorship, adult insurance coverage, and disability benefits early in the process, whether or not the patient is ready to participate and fully understand these topics. These conversations should not be put off until the year or two prior to transition. Assessment of and action in these areas should be addressed early and often in the transition process [17].

Logistically, the clinic office managing a transition will need digital or paper files containing assessments, checklists, legal and insurance documentation, and medical summaries. It may be beneficial to provide a set of this information for patients as well to allow them to track their own progress and gain skills in self-management. Clinics may benefit from a database of all patients involved in the transition process to allow for rapid, efficient check-ins on clinic-wide progress and needs. Some clinics may also benefit from dedicated transition clinic days where multidisciplinary transition team efforts can be focused in a dedicated clinic environment. It is important for members of the transition care team to have clearly designated roles for specific areas of transition, such as selecting adult providers including subspecialists and making appointments, addressing education and vocational goals, managing insurance, etc., to reduce redundancy in clinical time and efforts. Documentation may be aided by formatting templates in the electronic medical record that can be edited and seen by all members of the team or paper or electronic tracking sheets shared by providers. At each routine visit during the transition period, progress should be evaluated and interventions tailored as needed to keep patients on track to transition between ages 18 and 22. It should be noted that not all patients fit this structure; depending on severity of illness and prognosis, some patients may be better served by remaining with their pediatric providers, particularly if life expectancy would not yield the patient experience to be best served by establishing new providers at the end of life.

Throughout the process, providers should make intentional efforts to include the patient directly in interviews and decision-making. This includes eye contact while speaking, directing questions to the patient, and interviewing independently for at least part of every visit. The goal of these adaptations to the classic parent-centered style of interviewing, prevalent in pediatrics, is to gradually empower the patient to take more ownership of their health and needs and to learn to speak for themselves. These skills will be invaluable in the adult care setting, as well as in efforts to maximally function in education, work, and social settings. Parents may need encouragement to allow their children to take over in this manner, and the provider is in an excellent place to set expectations and gently encourage graduated patient autonomy.

Several models of these assessments, checklists, medical summaries, and emergency plans are available from various transition support programs. One well-tailored to this population can be found in the Transition Toolkit for DMD supplementary materials: Readiness Assessment, Checklist, Medical Summary [39]. This resource has documents specifically adapted for patients with DMD, which will address the needs of many patients in the NMD world and provides a thorough outline for adaptations to patients with other neuromuscular disorders. Additionally, free sample documents are available to download from the Got Transition website. These are available in groupings specific to providers

facilitating pediatric to adult care transfer, the receiving adult providers, and those who will have continuity but need to adjust care as patients grow (family medicine and internal medicine-pediatrics). Documents are much more generic to suit the needs of patients with a whole host of conditions but can be adapted to specific patient needs. The Got Transition materials also have a wealth of information about developing a general transition clinic policy and process. Clinics that care for patients with chronic neuromuscular respiratory failure would be advised to create their own templates using the resources above, including the child neurology transition checklist example [17, 37].

In preparing patients to transfer to adult care and potentially into more independent environments in other areas of life, providers have a unique opportunity to outfit patients with an effective emergency plan. This can be helpful at any age or stage of illness but is particularly useful if the patient is away from primary caregivers and medical institutions. Patients should have easy, constant access to pertinent medical information at all times in case of emergency. This should include, at minimum, primary medical conditions, allergies, medications, and any emergency plans specific to their condition that may be unfamiliar to community providers. This information can be in a number of formats – a pocket card with relevant medical details, a phone application (e.g., Medical ID is already standard on iPhone or Android), or a digital copy of a medical summary on phone or in email. The Got Transition website and Toolkit for DMD supplementary documents provide excellent guidance for what to include. There is also an emergency pocket card available from the Got Transition resources page. Providers should also address the hospital of choice at the patient's primary location (may change with school, moving) [17].

Once the patient has transitioned into the adult clinic setting, referring and accepting providers have an excellent opportunity to collect feedback and outcomes data to guide improvements to the transition process. Phone interviews, written or online evaluations, and dedicated conversations during clinic visits can yield invaluable insight into the patient and family experience of the transition process and inform providers on areas for improvement and success. Objective outcomes data can also provide much needed evidence to the field of transition medicine, in general, and to this population in neuromuscular disorders, specifically. Several different areas of medicine have small studies of parent-patient-provider feedback in the transition process, which may be helpful in formulating new studies specific to NMD [28, 32].

Potentially Helpful Resources

- People-Process-Performance from MDA, https://strongly.mda.org/the-three-ps-of-transitioning-from-pediatric-to-adult-clinical-care/
- https://gottransition.org/providers/index.cfm
- www.childneurologyfoundation.org/transitions

There are many different models in the world of transition medicine, which are ever-changing and adapting to the needs of patients and institutions. Depending on the strengths and needs of a population or institution, providers could consider the following structures as outlines for their own programs:

- Inpatient transition team or consult service. This structure may include an inpatient care team dedicated to patients in the transition age group and attend to their needs with transition planning in mind. The consult service model could allow providers familiar with the process to meet patients during an inpatient stay and help them organize a plan to meet their needs in transitioning providers and potentially institutions.
- Dedicated transition clinic, specifically to address transition readiness and planning and follow patients in the pre- and post-transition phases of their care to provide some continuity in the process. This could be disease specific or available to any AYA with medical complexity.
- Creating a day or half day specifically set aside for patients in the ages of transition preparation or action, which could be specific in a disease process (i.e., spina bifida clinic or Duchenne clinic) or a general location with support staff to allow various specialties and conditions to utilize the environment (i.e., Tuesday PM genetics, Wednesday AM NMD, Friday PM DM1).
- A planned follow-up visit or contact (email, phone, etc.) after the first appointment(s) with adult providers to assess for additional needs or areas for improvement in the process (i.e., Do you feel comfortable with your new provider? How could we make the process better?)
- Establish a continuity provider who will remain with the patient through the transition to adult subspecialists and is comfortable and able to assist in this process (i.e., primary care provider in family medicine or internal medicine-pediatrics, social worker, cross-trained respiratory therapist).
- Identify a provider in your group who will handle all of the outgoing or incoming transition patients in your practice. This could consolidate resources and experience in a narrower set of clinic staff.

Summary with Areas for Progress

Over the past couple of decades, substantial advancements in disease-specific therapies and chronic condition management have drastically changed the patient experience with many NMDs. This has created incredible opportunities for patients and families, as well as new frontiers and challenges for the medical teams caring for these patients. There is ample opportunity for research in many different areas, particularly with regard to quality improvement in provider education and the transition process [3, 41]. We have minimal data to guide interventions; any contributions of research, care models, treatment strategies, or institutional policy can be helpful

in cultivating best practices in the ever-changing care of patients with NMDs. Continuing medical education has a vital role to play on both pediatric and adult sides, but special focus on augmenting education and training in these previously pediatric-specific disease processes for adult-trained providers is warranted [22, 40]. As a medical system, we have a prime opportunity to work to improve the outcomes and experiences of AYA with chronic health needs [16].

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