

MEDICAL SCHOOL HOTLINE

Medical Student Insights on Pediatric Neuromuscular Disease in Hawai'i

Nina L. Beckwith MEdT; Daniel W.W. Hong BA; Kelsey Y. Ige BA; Keli Y. Tahara BS; and J. Douglas Miles MD, PhD

The Medical School Hotline is a monthly column from the John A. Burns School of Medicine and is edited by Satoru Izutsu PhD; HJMPH Contributing Editor. Dr. Izutsu is the vice-dean of the University of Hawai'i John A. Burns School of Medicine and has been the Medical School Hotline editor since 1993.

Background

The Muscular Dystrophy Association (MDA) is an organization that provides services to people with certain debilitating nerve and muscle diseases and their families. There are 43 such diseases that MDA includes in its mission, including Duchenne and Becker Muscular Dystrophies, the various Charcot-Marie-Tooth disease subtypes, myasthenia gravis, and various motor neuron diseases. Hawai'i's local chapter, MDA Hawai'i, estimates that about 500 families in Hawai'i are currently affected by the neuromuscular diseases under their purview.¹

MDA Hawai'i organizes and conducts an annual Summer Camp for children ages six to seventeen years living with various neuromuscular diseases. These summer camps are held at Camp Erdman on the North Shore of O'ahu. At these week-long camps, the children, who have varying degrees of disability, participate in activities such as swimming at the pool, singing songs around a campfire, dancing at formals, performing at the talent show, and creating arts and crafts. This is an opportunity to make friends with peers and spend time away from home and parents.

To ensure the safety of these children, many of whom are medically fragile, MDA recruits a large number of volunteers to spend the week with them. Most volunteers serve as camp counselors. They also assist with the campers' daily needs, which may include medical needs and assistance with activities of daily living (showering, dressing, and toileting).

For the first time, students at the John A. Burns School of Medicine (JABSOM) enrolled in a summer elective that involved volunteering at the MDA Hawai'i Summer Camp. JABSOM requires all students to engage in a course titled "Explorations in Medicine," or simply "MD5," during four of the eight weeks between the first and second year of medical school. In this course, students are encouraged to explore their interests, "whether it is learning more deeply in the biological sciences, shadowing a physician in a clinical setting, or conducting research."² Some students travel to clinics abroad while others enjoy opportunities closer to home. In July 2015, four second-year medical students volunteered as camp counselors at the MDA Summer Camp. They lived with their campers for the

entire week and became their friends and caregivers. Vignettes and reflections on the camp experience are explored below.

Textbooks vs Experience

Despite profound weakness in his neck and limbs, my camper discovered a style of swimming that was not only comfortable for him, but which allowed him to interact fully and play with the other campers. While in the pool, he reminded me that I would act as his arms and legs when he and the other campers played "sea monster," a game requiring the use of one's extremities.

MDA Summer Camp was an opportunity for the medical students to look at neuromuscular disease from two different perspectives: as medical students and as camp counselors. As medical students, they gained an understanding of the pathophysiology underlying neuromuscular disease. As camp counselors, they gained insight into how the clinical manifestations of the diseases affected the children as individuals.

The medical students not only observed physical signs and symptoms of neuromuscular disease, but began to understand what those manifestations meant to the patient. In medical school, the approach to the study of disease is structured by organ systems and allows students to broadly cover a variety of common diseases. The focus is directed at the mechanisms involved in producing disease with less emphasis on the implications of the disease on an individual's health and well-being. Even when those implications are confronted, they are addressed in an academic, non-clinical setting. Reading about fatigue related to muscle disease conveys a certain level of understanding, but bonding with someone who is experiencing that fatigue conveys a different, deeper understanding. The MDA Summer Camp allowed the students to connect the clinical aspects of diseases to specific consequences, such as requiring head support while swimming due to profound neck weakness.

Phenotypic Variability

Two siblings with the same inherited myopathy presented differently from one another. One eagerly jumped off diving boards and ran tirelessly when playing with his peers, while the other

found it difficult to hold her breath underwater and required a wheelchair and BiPAP when she overexerted herself. Her fatigue largely varied from day to day; one day she requested to sit in her wheelchair and could not tolerate walking short distances, while on other days, she could dance for hours with minimal breaks after being very active earlier that day.

Many medical texts may generalize disease processes based on their typical clinical presentation, offering only a snapshot of the disease. However, diseases do not always manifest exactly the way they are described in textbooks. There can be considerable variability in the phenotype of a disease, as demonstrated by the vignette above. The students recognized how muscle weakness and fatigue fluctuated from day to day in a single individual. They also noted the variability in the degree of these symptoms among individuals with the *same* condition. The students found it more appropriate to understand symptoms along a spectrum.

Perception of Neuromuscular Disease

The first time I took her out of her wheelchair, I sat her on the bed and started to lift her legs, thinking that she needed assistance positioning herself in bed. However, she began reaching for her knees, so I retracted my hands and observed as she maneuvered her legs, bringing them onto the bed using the strength in her arms. Lying down, she carefully used the wooden beams of the bunk bed to roll herself from one side to the other, and she used her hands to move her legs around the bed. At that moment, I realized she was not as fragile as I thought and perhaps I was doing too much to help her during her daily activities.

The challenge in working with medically fragile children is finding a balance between being helpful and being patronizing when considering the campers' abilities. As in the story above, counselors may underestimate the abilities of their campers and feel inclined to offer more assistance than necessary. However, many of the children did not perceive their condition as limiting. One camper, for example, had severe muscle weakness in his lower extremities and was unable to stand on his own. Despite these "limitations," he participated in activities that traditionally require the use of leg muscles, such as swimming, jumping off a diving board, and dancing.

While there were campers who refused to be restricted by their conditions, others were dissuaded from attempting new activities out of fear of triggering symptoms. One camper wished to swim at the deeper end of the pool, which would require her to pass a swim test. However, because of her limited pulmonary function, holding her breath for even a short period of time could trigger a headache. Thus, she elected not to take the swim test. While many campers refused to be limited by their conditions, some were reluctant to try new activities out of fear of worsening their symptoms.

In addition to being aware of the way counselors perceived their campers and the way the campers perceived themselves, the students also noted how the campers were viewed by other children. In general, people seem unsure of how to interact

with individuals who are physically different from themselves. Devices such as wheelchairs or physical features such as scars can create barriers to interaction. All campers at Camp Erdman, including those from MDA and other summer camps, shared the same dining hall. Other campers frequently stared at the MDA campers. For campers with more serious symptoms, the psychosocial barriers were apparent. For example, one camper was only able to move her eyes and needed medical equipment with her at all times. Many campers and counselors did not know she could communicate with eye movements. Few people other than her caregivers stopped to talk with her, perhaps largely due to the false assumption that she was unable to communicate. Learning how to care for medically fragile children at the MDA Summer Camp helped the students recognize and overcome certain barriers between caregivers and patients, which will be beneficial for future clinical encounters.

Children as Advocates of Their Own Health

A camper with muscle weakness and reduced pulmonary function fatigued easily from physical exertion. During the dance, she paused to take a break and instinctively reached for her heart rate monitor. She explained to me that she used it to gauge whether or not she needed to go on her ventilator. Aware of her energy level and physical state, she also spoke about needing to ride in her wheelchair when she felt tired throughout the day. Because she was able to identify and express her needs, caring for her was a relatively simple task.

Most of the camp volunteers had limited experience in caring for individuals with neuromuscular diseases. Assisting medically fragile children with daily activities, such as bathing, feeding, and operating medical equipment were new tasks for the volunteers. This unfamiliarity compelled campers to communicate their needs and preferences. As demonstrated above, to ensure effective care, these children needed to communicate fluctuations in their health. When campers took the initiative to voice their opinions and preferences throughout the week, it was easier for their counselors to care for them. Advocating for their own health is a lifelong skill for these children, one that ensures optimal care and improves quality of life.

Parents play a vital role in teaching their children to be advocates of their own health. For instance, one camper usually takes a medication in powdered form, without any food or liquid. At camp, the medical staff, unaware of her preferences, mixed her medication with pudding or applesauce. After camp, her mother informed the counselor that her daughter prefers to receive her medication as a powder. The mother reminded her daughter to voice her preferences about her medication and other aspects of her health. She reinforced the idea of the importance of being a "leader in her personal care."

Empathy

I first met my camper in the afternoon. As I pushed her wheelchair around camp, I was very conscious of the bumps in the road, trying to maintain my eyes on her eyes, and trying not to

juggle all the medical equipment attached to her wheelchair around too much. But little did I know, the middle of her day was the easiest part--she started her mornings with an hour and a half of routine treatments, and she ended her day with the same care.

Some clinicians would argue that empathy – the ability to understand and be sensitive to the experiences of others – is an important part of patient interaction.³ In addition to making the clinical encounter more pleasant for the patient and helping them feel better cared for, heard, and understood, empathy also helps clinicians begin to understand how patients experience their disease, understand their limitations, and adjust accordingly.

Prior to camp, the medical students were unfamiliar with neuromuscular diseases, and despite reading about them in textbooks, the most they could do was sympathize with their future campers. Living with the campers at the MDA Camp gave the students an opportunity to speak with the campers about how their conditions affected their everyday lives and what the diagnostic process was like from their perspective. It is through clinical experiences like the MDA Camp that students may begin familiarizing themselves with such conditions and thus work towards developing empathy.

Medically fragile children and others living with chronic conditions require a strong support system to ensure their health and wellbeing. The medical students gained an appreciation for the value of having a team approach to care and the importance of being mindful of the psychosocial impact of disease on the individual and those involved in their care. Through assisting campers with daily activities, the students realized how demanding these illnesses can be for parents. In addition to providing physical care, the students recognized that families also play a role in helping their children psychologically cope with the physical manifestations of their conditions. These are not easy feats, and doing this on a regular basis can be physically and emotionally draining for parents and caregivers. Thus, it is important for health care providers to be sensitive to the reality of burnout in caregivers and the potential strain of one's medical condition on relationships. Any of these can quickly create environments that are unfavorable for the child.

Conclusion

Learning about diseases from texts differs from the practical experience of spending time with someone who lives with the disease. Textbook descriptions of disease may also differ from the actual appearance of the disease when seen firsthand, and disease may present with considerable phenotypic variability that cannot be fully appreciated from didactic learning. Furthermore, no one can fully understand the lifetime of hardships a patient will experience or has already encountered by simply talking to them. Even though the students caught a glimpse of what life is like with neuromuscular disease, this experience has made them more sensitive to the various ways neuromuscular

diseases may impact children and their families. Hopefully, this awareness will better equip them to understand the struggles and anticipate the needs of similar individuals or families, whether those needs may be medical, physical, occupational, or psychosocial.

Educational Perspective

This opportunity was created as a four-week elective for JABSOM students who had just finished their first year of medical school. During the first week of the rotation, the students received didactic teaching on nerve and muscle physiology, neuromuscular diseases, and the neurological physical exam, with an emphasis on findings pertinent to peripheral nervous system disorders. They were given reading assignments on neuromuscular disease in general and on specific diseases including the dystrophinopathies, spinal muscular atrophy, Charcot-Marie-Tooth, and other myopathies. They also shadowed a neuromuscular neurologist at his clinic. The second week, they served as volunteer counselors at the MDA Camp. For the remainder of the course, they continued to shadow in the neuromuscular clinic and wrote up reflections on their experience.

From an educational perspective, the medical students found it surprising that much of their experience was learning about the psychosocial aspect of the neuromuscular disease when they were expecting to learn more about the pathophysiology or clinical aspects of the disease. Half of the medical students felt this experience helped with their understanding of the basic science underlying the disease, while half did not. There was a consensus that the experience would help them clinically. However, all four students considered the experience to be of educational value and stated that they would recommend the experience to other medical students.

The rotation provided exposure to neurological and pediatric issues. It allowed the students to see first-hand a collection of rare diseases that otherwise would not likely be encountered during the course of medical school.

MDA Perspective

The experience was not only beneficial to the medical students, but also for the campers and for MDA. Adrianna O'Donnell, former executive director of MDA Hawai'i, commented on the involvement of JABSOM medical students in the MDA Summer Camp by indicating that their involvement was a valuable addition to the camp. She plans to repeat this opportunity for medical student participation in future years.

Ms. O'Donnell described the medical students as "organized, well-spoken, responsible and respectful. They were genuinely interested in our campers, their condition and how to care for them best."¹ She added, "Our volunteer counselors are the reason camp is so successful. They provide care, fun, friendship and support in a trusting environment. Med students not only demonstrated integrity and intelligence, but they were also able to connect with their campers as friends."¹

Summary

The experience at the MDA Camp reinforced the importance of adopting a patient-centered approach to healthcare that is individualized and adaptable to patient needs through time and circumstance. Engaging in daily activities with pediatric patients affected by neuromuscular diseases can provide deeper understanding of the various limitations patients face, of how they adapt, and of their perceptions of their diseases. This in turn can provide insight into how to best approach patients in the clinical setting to optimize their health and quality of life.

This experience was also a lesson in working with individuals with neuromuscular disease. This subpopulation of pediatric patients is a unique one, with specific needs that may be difficult for health care providers to approach. Learning how to interact with these patients and to ask the appropriate questions comes with experience, and volunteering at the MDA camp provided an opportunity for growth.

The medical students found it surprising that much of their experience was learning about the psychosocial aspect of the neuromuscular disease when they were expecting to learn more about the clinical aspects of the disease. Medical education teaches students the facts and figures related to the pathophysiology of disease, and it introduces them to management algorithms in disease treatment. This is necessary, given the vast amount of material one needs to learn in four short years. However, there are still some experiences students and physicians can participate in – even in the context of clinical education – that can help remind physicians of the human elements of our profession. These experiences help strengthen the more intangible skills of medical practice in ways that classroom learning cannot. In this case, all participants—campers, MDA Hawai‘i, and the students—benefitted from the collaborative effort.

Authors' Affiliations:

- Second-year medical students, University of Hawai‘i John A. Burns School of Medicine (NLB, DWWH, KYI, KYT)
- Assistant Professor, Department of Medicine, University of Hawai‘i John A. Burns School of Medicine (JDM)

References

1. Personal correspondence with Adrianna O'Donnell, executive director of Muscular Dystrophy Association Hawai‘i, July 30, 2015.
2. Horio D. MD5: Explorations in medicine: handbook for a course at the University of Hawai‘i John A. Burns School of Medicine. 2014 (unpublished).
3. Jani BD, Blane DN, Mercer SW. The role of empathy in therapy and the physician-patient relationship. *Forsch Komplementmed.* 2012;19:252-257. doi: 10.1159/000342998.