



An Exploratory Case Study to Support Young Children with Spinal Muscular Atrophy (SMA)

Sheng Miao, Ziyang Tang, Jinjuan Heidi Feng
Computer and Information Sciences Department
Towson University
Towson, MD21252, USA
{smiao1, ztang, jfeng@towson.edu}

Amanda Jozkowski, Molly Lichtenwalner
Department of Occupational Therapy
and Occupational Sciences
Towson University
Towson, MD21252, USA
{ajozkowski, mlicht4@towson.edu}

ABSTRACT

In this paper, we describe a preliminary case study that examines the challenges faced by very young children with Type I Spinal Muscular Atrophy (SMA) and how technology may help these children live a more independent life. Several input solutions were examined to support interaction between a young patient and computer-based systems. We started working with the patient when he was two and a half years old. The challenges observed and lessons learned regarding both working with very young children with severe disabilities and the use of specific technical solutions are discussed.

CCS Concepts

• Human-centered computing → Accessibility → Empirical studies in accessibility • Accessibility technology

Keywords

Young children, Spinal Muscular Atrophy (SMA); Sensor

1. INTRODUCTION

Spinal Muscular Atrophy (SMA) is an autosomal recessive neurodegenerative disease characterized by degeneration of spinal cord motor neurons, atrophy of skeletal muscles, and generalized weakness [1]. The incidence of SMA is about one in every 6,000 to 10,000 live births with a carrier frequency of one in 50 [2]. Nowadays, SMA affects more than 750,000 patients worldwide and is the number one genetic killer of infants and toddlers [3]. SMA affects an individual's motor neurons in a range of ways depending on the particular type; people with SMA are often unable to walk, many cannot sit up unassisted, and (in the most severe forms) are unable to eat and breathe. Although researchers have identified the gene that causes SMA, and potential parents can be screened to find out if they are a carrier, there is currently no known treatment or cure [1].

2. CHALLENGES AND OBJECTIVES

Our research team worked together with a very young patient Kevin¹ with Type I SMA and his family since Kevin was two and a half years old. While Kevin is fully intact cognitively, he is

¹ The participant's name has been changed to protect anonymity

unable to function independently in many domains. He uses a tracheostomy tube and ventilator to breathe, and requires assistance to sit and to hold his head up. He wears an exoskeleton and other assistive/mobility devices to help him utilize his minimal movements (due to extreme muscle weakness), and must have 24-hour supervision to monitor his vital signs. Despite these challenges, he and his family, together with a large and dedicated team of physicians and therapists, have made incredible strides toward making his life as "typical" as possible. He enjoys looking at books, listening and dancing to music, watching his pet fish, playing with playdough and puzzles, and going outside to the park and the zoo.

One area that Kevin's parents are struggling with is helping him communicate and control his environment as desired. His parents spoke about his intelligence and obvious desire to interact and engage with the world, but they felt that he was completely dependent on others for all aspects of his care and participation. They envision Kevin as being able to independently participate – for example he might wake up, call for his mother, turn on the lights in his bedroom, and then select and start playing a computer game while he was waiting to be fed. Currently, to perform these tasks, Kevin would need someone to notice he was up, turn on the lights for him, ask him what he wanted, set up a communication device or attempt to interpret his vocalizations (he is unable to form words due to low facial muscle tone), bring over a switch-adapted toy or computer tablet, hook up his hands and position everything correctly, and then stay with Kevin to make sure everything is in place and usable. Kevin is not capable of repositioning devices by himself if they are moved. In addition, many of the products currently available require too much force for Kevin and other children with Type I SMA to actuate, or they are not as sensitive and accurate as needed for the desired applications.

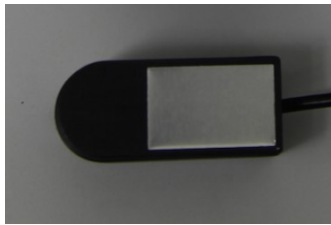
We believe that Kevin has the potential and the underlying capacities to be able to operate devices such as an eye-gaze communication system, a power wheelchair, a personal environmental control system, etc. if appropriate input devices are identified to fit his unique constellation of needs. The following section will discuss one specific aspect of our project: the design and preliminary evaluation of the input solutions.

3. TENTATIVE INPUT SOLUTIONS

While building the communication and environment control systems for Kevin, one of the main challenges is the channel of input due to his extremely limited physical and vocal capabilities. Eye-gaze based solution had been evaluated and yielded promising results. But the eye-gaze based solution can be intrusive in the context of communication. Regarding hand

controlled input solutions, the following characteristics were identified based on parents' input and observations:

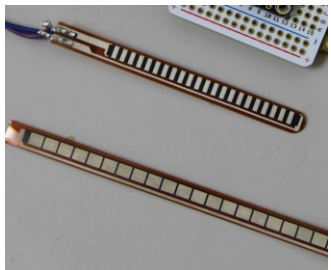
- Customizable (i.e., can be modified/adapted for others with Type I SMA and similar disorders)
- Affordable/accessible to people in the SMA community
- Usable (does not take more than 5 minutes to set up and can be easily understood and modified by families who are not technically trained)
- Durable
- Responsive/sensitive
- Accurate
- Allows for maximal functional movement patterns (i.e., palms of hands are not obstructed so they can still be used for play)



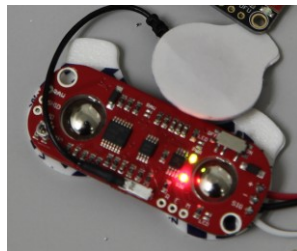
(1a)



(1b)



(1c)



(1d)

Figure 1. Demonstration of four input solutions: (a) micro light switch; (b) conductive fabric thread; (c) flexible sensor; (d) EMG-based muscle sensor

Based on the above listed criteria, we chose and tested four types of input techniques as demonstrated in Figure 1. Figure 1a illustrates a micro light switch that we positioned under Kevin's index and middle fingers. Figure 1b illustrates a finger cot that we knitted for the size of Kevin's hands using conductive fabric thread. Figure 1c illustrates two flexible sensor with different lengths. Figure 1d illustrates an EMG-based muscle sensor.



Figure 2. Demonstration of the flexible sensor solution during testing

Figure 2 shows a customized approach to attach a flexible sensor on to a glove specifically made for Kevin's hands to better capture the intended movements.

4. RESULTS AND CONCLUSIONS

A preliminary evaluation of the solutions suggests that the micro light switch might be the solution that offers a reasonable balance between efficiency and accuracy. Several challenges and design considerations for the targeted population have also been observed. First, substantial amount of the hand movements are passive and it is very difficult to differentiate passive movements from intentional movements. Second, the threshold for movement detection needs to be set very carefully. High threshold may make it too hard for the patient to generate detectable movement, while low threshold may make the system too sensitive and incapable of filtering out noises in the movements. We also found that the thresholds can be quite different between the two hands if the muscle tone of one hand is stronger than the other. Third, collecting empirical data from very young children with severe disabilities takes much longer time and preparation compared to neurotypical adult users. The tasks must be age appropriate and highly engaging. In this study, we developed an interactive game based on Kevin's favorite character, the minions, to evaluate the input solutions. Even with this customized game, Kevin had often times lost interest in the middle of a task and refused to continue.

Currently, we are recruiting more young children with SMA to evaluate the input solutions. Like many health conditions, the capabilities of young children with SMA vary substantially. Data from more target users will help better understand how to identify and customize effective solution(s) for a particular user.

5. ACKNOWLEDGMENTS

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6. REFERENCES

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