School of Biomedical Engineering, Science, and Health Systems

Design and Testing of a Functional Arm Orthosis in Patients With Neuromuscular Diseases

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***Abstract*—The objective of this study was to determine the utility of a passive gravity-balanced arm orthosis, the Wilmington robotic exoskeleton (WREX), for patients with neuromuscular diseases. The WREX, a four-degrees-of-freedom functional orthosis, is energized by rubber bands to eliminate gravity and is attached to the wheelchair. The development and clinical testing of WREX is described in this report. Seventeen patients (14 boys and 3 girls) with muscular disabilities participated in the study. Ages ranged from 4 to 20 years. Criteria for inclusion included a weakened arm, use of a wheelchair, the ability to grasp and release objects, and the ability to provide feedback on device use. Testing consisted of administering the Jebsen test of hand function without WREX and then testing again after approximately two weeks of wearing the WREX orthosis. The timed results of each task within the test then were compared. Specific tasks related to vertical movement required less time to perform with the WREX. A large number of subjects were able to perform the Jebsen tasks with the WREX, where they were unable to perform the task without the WREX. Patients can benefit from WREX because it increases their performance in daily living activities and makes many tasks possible. The range-of-motion in the patients' arms increased considerably, while the time required to complete some of the Jebsen test tasks decreased. Most patients were very receptive to WREX, although a few were ambivalent.**

***Index Terms*—Arm orthosis, muscle weakness, neuromuscular disease.**

I. Introduction

MUSCULAR DYSTROPHY (MD) and spinal muscular atrophy (SMA) are two of the more common neuromuscular diseases that affect children. These diseases produce systemic muscular weakness, which creates a sense of frustration and futility that often accompanies significant dependency on others for personal care. The goal of this project was to reduce this dependency by developing a mechanical arm orthosis to enable users to complete many tasks independently.

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Duchenne muscular dystrophy (DMD), the most common form of muscular dystrophy, occurs when there is a genetic failure to produce the protein dystrophin or there is a defect in the gene that produces dystrophin. Duchenne muscular dystrophy occurs in approximately one of every 3500 live male births [1]. Becker muscular dystrophy (BMD), another form of MD, tends to be less severe than DMD and is much less prevalent. It occurs when the amount of dystrophin produced is insufficient or poor in quality. Congenital muscular dystrophy (CMD) defines a group of disorders in which children are weak at birth or become weak within the first few months of life and have specific dystrophic changes in muscle biopsy. Congenital muscular dystrophy, in general, is relatively non-progressive, but in some cases, progression is rapid. Children in this study had one or more of these conditions.

Brussock *et al.* [2] compared the strength of different muscle groups of children with and without DMD. They used an electronic force sensor to measure isometric force strength in children ages 4 to 16 years with DMD. They also tested age-matched controls. The researchers showed that isometric force ranged from 34.3 to 174.6 N for normal patients, while force for patients with DMD ranged from 4.9 to 57.9 N. Because of this weakness, the ability to perform activities of daily living decreased over time, and the individuals increasingly relied on caregivers.

The other group of children in this study had SMA, which refers to a group of inherited diseases characterized by muscle wasting and weakness. It affects one in every 10 000 live births [3], [4]. It is an autosomal recessive hereditary disease caused by a deletion in a portion of the survival motor neuron gene. There are three types of SMA: Werdnig–Hoffman (Type I), intermediate (Type II), and Kugelberg–Welander (Type III). Children diagnosed with Type I are very weak and typically have difficulty sitting unsupported. Type II is less severe: children are able to sit without support although they usually need help getting into the sitting position. They are able to stand with the assistance of braces. Type III is the mildest form; individuals can stand and walk alone although perhaps with some difficulty. Patients in the current study had Type II SMA. Spinal muscular atrophy, like MD, affects the larger muscles closer to the body rather than those at the extremities. As in other forms of neuromuscular disease, decreased movement of an extremity over time leads to contractures and a decrease in the range-of-motion (ROM). Wang *et al.* [5] emphasized the need for ROM exercises in the upper extremity, particularly elevation and abduction, for patients with neuromuscular disease.

Early research in upper-limb orthoses was performed at the Case Institute of Technology in the early 1960s [6]. A powered exoskeleton was connected to the floor and controlled by a light source mounted on the wearer's head that activated light sensors in the environment. This device could be moved either directly or be preprogrammed to complete different tasks. This research led to the development of the Rancho "golden" arm at Rancho Los Amigos Hospital, Downey, CA, in 1969 [7]. This device was a powered orthosis with seven independent joints and was controlled with tongue switches. Development has continued at the Rancho Los Amigos Hospital with the development of the Rancho-JAECO mobile arm support, which is an improved version of the mobile arm support [8].

Other orthosis projects include Engen's pneumatic orthosis system [9], the Burke orthosis [10], the Musgrave orthosis [11], an exoskeletal mobilizer for amyotrophic lateral sclerosis [12], an intelligent rehabilitative orthotic system [13], an upper-limb motion assist system [14], and a motorized upper limb orthotic system [15]. These systems generally were fairly impractical or aesthetically unappealing. None saw commercial success. One project of note is by Herder *et al.* [16], who have developed an elegant arm support device with springs mounted in the base. Clinical trials are ongoing in this project.

The most popular orthosis used today is the balanced forearm orthosis (BFO) [17], a body-powered device developed in the 1950s. The BFO has three joints and allows the person to move in the horizontal plane. It has a pivot at the forearm that allows patients limited ability to move their hand up and down. However, this device does not provide 3-D movement, and it is difficult and time-consuming to fit. There is a vertical-movement version of the device that uses elastic bands, but it is only in equilibrium at one vertical level and is seldom used by patients.

The current study reports on a body-powered orthosis design that is modular and mounted to a person's wheelchair. The Wilmington robotic exoskeleton (WREX) is a two-segment, four-degrees-of-freedom exoskeletal arm, energized by elastic bands that aid in moving the arm in 3-D space. The WREX allows full passive ROM of the arm and provides a sense of flotation that assists in voluntary movement [18].

There are very few functional devices available to people with upper-limb weakness that can restore some of the lost motor capabilities in the hand. Conversely, amputees have a number of passive and active prostheses available. This area has seen considerable research over the years, beginning with the post-WWII era. Apart from the BFO and various overhead slings, no arm-assistance devices exist. This project attempts to fill this void by developing and testing a simple, low-cost device that provides 3-D movement and can be configured easily by a rehabilitation technician or occupational therapist.

II. Methods

A. Participants

Upon Institutional Review Board approval, 17 patients (14 boys and 3 girls) with neuromuscular disabilities were enrolled in the study. Seven patients had DMD, one had BMD, four had SMA, and five had CMD (Table I).

All were right-handed. Ages ranged from 4 to 20 years. Study inclusion required arm strength of less than 5 on the manual muscle test (MMT) scale [19]. The MMT evaluates the function and strength of different muscles and muscle groups based on the ability to move in relation to the forces of gravity and manual resistance. The test grades the strength of the muscles from 0 (trace- no visible or palpable contractions) to 5 (normal). An additional requirement for study inclusion was regular use of a wheelchair to which the orthosis was affixed. All but one subject used a powered wheelchair.

TABLE I
Patient Data

Patient	Age (yrs)	Sex	Weight (Kgs)	Disease	MMT strength		
					grip	deltoid	bicep
1	16	M	80	SMA	-	-	3 to 3-
2	12	F	46	CMD	-	3	4
3	11	M	79	DMD	5	3	4
4	11	M	29	SMA	4	tr	3-
5	16	M	61	DMD	5-	3-	3 to 3-
6	17	M	40	DMD	4	2+	2+ to 3-
7	12	M	82	DMD	4	3+ to 4-	-
8	14	M	60	DMD	4	3	-
9	15	M	69	DMD	3	tr	3-
10	18	M	68	DMD	4	tr	tr
11	11	M	23	CMD	3+	3+	3+
12	13	M	29	CMD	2	3-	4-
13	4	M	13	SMA	-	3	3
14	9	F	47	SMA	3	tr	3-
15	8	M	24	CMD	-	4	4
16	14	F	51	CMD	3+	tr	3
17	20	M	78	BMD	4	2+ to 3-	3

SMA = spinal muscular atrophy, CMD = congenital muscular dystrophy, DMD = Duchenne muscular dystrophy, BMD = Becker's muscular dystrophy, MMT = manual muscle test, tr = trace

All patients had difficulty moving their arms. This was most evident in feeding, which was difficult for most of the participants. The patients routinely used compensatory techniques such as assistance from the contralateral hand, "crawling" up their body, hooking a finger into their mouths to position their elbow to get better leverage, and using a tray to assist in elevation. Sixteen patients were fed orally; one was tube-fed. None used an assistive device, such as a BFO, for arm movement prior to the study. Patients were recruited through the muscle clinic at the hospital. After receiving a description of the project, the patients read and signed the informed consent documents, which were prepared and administered in accordance with Institutional Review Board guidelines. For the evaluation phase, the Jebsen test of hand function, which required patients to have the ability to grasp and release objects, was administered. Also, each participant completed a survey about the orthosis and its use. Data for all patients are listed in Table I.

B. Apparatus

The WREX system for the upper extremity was designed and built at A.I. duPont Hospital for Children. Detailed design information can be found in Rahman *et al.* [20], [18]. The goal of the WREX is to provide a sense of flotation that encourages a person with neuromuscular weakness to move his or her arms. Gravity-balancing the entire arm for all positions in 3-D space allows the patient to move the arm with very little effort. To negate the effects of gravity on the upper extremity and to achieve static balance, either counterweights or springs (or elastic bands) can be used to hold up the arm. The counterweights provide a balanced system for all positions but add weight and inertia and increase the size of the device. Springs

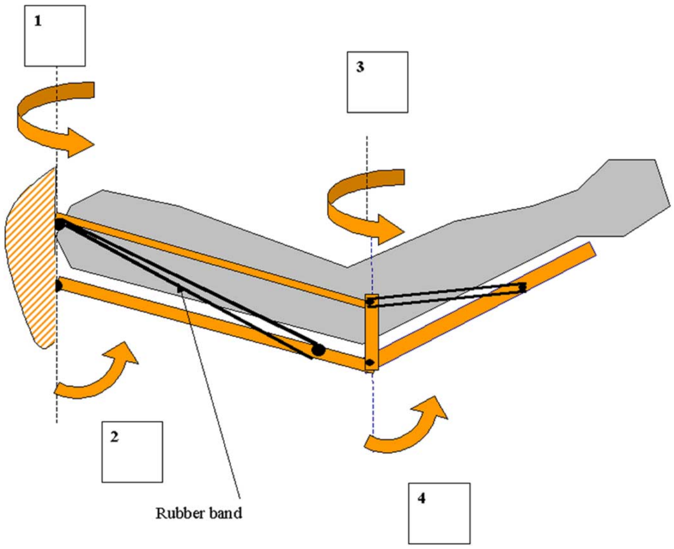

Fig. 1. Configuration of WREX showing the four-degrees-of-freedom. Upper arm utilizes a parallelogram structure, while the forearm has a single link to which is attached an arm trough. Entire linkage has four joints set up in the following order: 1, 2—shoulder joints; 3, 4—elbow joints.

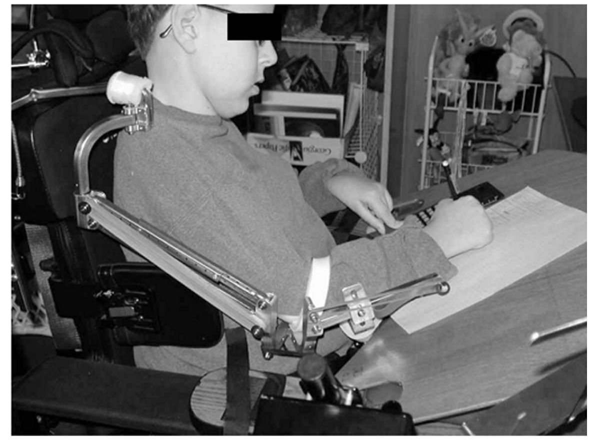

Fig. 2. Subject with WREX.

are more appealing because they store elastic energy and do not add appreciable inertia. This results in a more compact device.

The torques required at the shoulder and elbow to elevate the arm are nonlinear (there is much less effort involved in holding the arm straight up versus straight out in front due to the moment arm that varies in a nonlinear manner), and since the goal is to have a perfectly balanced system, it is necessary to introduce a nonlinear restoring force. This can be accomplished in one of two ways: 1) using nonlinear springs, or 2) using linear elastic elements such as bungee cords, therabands, elastic bands, or springs and then creating a nonlinear moment through geometry. Custom nonlinear coil springs are difficult to construct or purchase, and the result is not sufficiently compact. Linear off-the-shelf elastic elements (Sammons Preston, Cedarburg, WI) can be used judiciously to obtain a nonlinear torque through geometrical variation of the moment arm [20], [18].

The joint configuration for WREX is divided into two rotations at the shoulder and two rotations at the elbow (Fig. 1). This allows positioning of the elbow and hand in 3-D space but does not allow for pure axial rotation about the humerus or pronation/supination about the elbow. The two perpendicular joints at the shoulder combine to allow abduction–adduction and flexion–extension. The two elbow joints combine to allow elbow flexion–extension and partially compensate for the absence of humeral rotation at the shoulder.

The structure of WREX consists of a two-segment exoskeleton that shadows the upper arm and forearm (Fig. 2). The links are made from hollow steel rods and arranged in the shape of a parallelogram for the upper arm and a single link for the forearm. The upper arm is telescopic so the length can be adjusted to the size of the patient. The parallelogram structure is necessary so that the elbow joint remains vertical as the elbow moves up and down [20]. This vertical position is required to provide complete gravity compensation for both limb segments, for this particular configuration. The entire device is pivoted just above the shoulder. An arm trough is attached to the forearm link for forearm placement. The trough is custom-made for each wearer by casting the arm with plaster, fabricating a positive mold, and creating a negative polyethylene brace. Adding or subtracting the number of elastic bands accommodates individuals of different weights. These bands have a consistent visco-elastic behavior, are available in different levels of stiffness, and are easily identified by color.

The gravity compensation technique used here is an approximation of the technique described in Rahman *et al.* [20]. This approximation does not require the need to use zero free-length springs, which would provide ideal gravity compensation and give a sense of flotation. In practice, factors such as inertia, passive tissue resistance and spasticity offer resistance to motion that masks the gains due to zero free-length springs. Whereas using zero free-length springs adds to the mechanical complexity and manufacturing costs.

C. Experimental Procedure

After a patient agreed to participate and met the inclusion criteria, his or her arm was cast to obtain a custom forearm brace. The casting was performed at the hospital prosthetics and orthotics facility. The brace was then attached to the WREX. The patient returned to the laboratory, and the WREX was attached to the wheelchair. The upper-arm link was adjusted in length to match the anatomical limb. The number of bands was adjusted so there was just enough lift for each patient. These adjustments allowed the patient to move his or her arm freely in 3-D space. The WREX was fixed to the wheelchair by positioning the first joint (Fig. 2) above the shoulder. This provided optimal movement of the arm. A hollow length of steel tubing was then bent between the WREX and a wheelchair clamp. This provided a robust and precise connection. Four WREX units were constructed and rotated between the 17 patients in the study.

For the first round of testing, patients completed the Jebsen test of hand function [21] without the WREX. After they had used the WREX in their home for two weeks, they retook the Jebsen test with the WREX.

The Jebsen test evaluated each individual's disability as well as the effectiveness of treatment. This test was created to evaluate broad aspects of hand function used in daily living activities

TABLE II
QUESTIONNAIRE GIVEN TO EACH PATIENT AFTER THE STUDY

1. Did the orthosis perform well?
2. What would you change about the orthosis?
3. How much did you use the device daily?
4. Did you like the way the orthosis looked, if not, why?
5. What tasks did the device help you do?
6. What tasks did it not help you do that you would have liked?
7. Would you like to have such a device?
8. Did the device make you tired?

TABLE III
AVERAGE TASK COMPLETION TIMES OF THE SEVEN JEBSEN TASKS WITH THE NUMBER OF PATIENTS COMPLETING EACH TASK

Jebsen Task	Number of subjects completing tasks		Completion Time (s)	
	Without WREX	With WREX	Without WREX	With WREX
Writing	9	9	19 (±7.3)	21.9 (±4.1)
Turning cards	11	13	11.4 (±3.5)	12.9 (±5.5)
Small objects	8	12	21.8 (±11.9)	16.2 (±7.5)
Feeding	5	9	28 (±6.9)	24 (±15.9)
Stacking	10	12	13 (±6.1)	11.3 (±5.2)
Light objects	9	11	11.9 (±4.9)	10.8 (±4.1)
Heavy objects	5	6	21.1 (±16.8)	15.5 (±11.8)

Completion time is mean ± SD.

and provides unbiased measurements of several standardized tasks against which each patient's results can be compared. The test includes seven subtests; each represents a different daily living activity. They are as follows: writing a six- to eight-word sentence, turning over five 3 in × 5 in cards, manipulating small common objects such as paper clips and bottle caps, simulated feeding, stacking checkers, lifting large light objects (empty soup cans), and lifting large heavy objects (500 g cans). The test was administered in the same manner to each patient. The patients were seated in a chair with a height of 18 in (48 cm) at a desk that was 30 in (76 cm) high. To accommodate children with wheelchairs of different heights, a variable-height table was designed. The times of each task were measured using a stopwatch.

Once the patients completed the first round of Jebsen testing, they were fitted with the WREX and given basic instructions for its operation. They used the WREX for approximately 1 h under supervision of the research staff and their parent or caregiver. Once they were comfortable with the operation, they took the WREX home for a two-week period. During these two weeks, they were asked to use it as much as possible. They could take it to school or elsewhere and were free to use it as they wished. Upon the patient's return, the Jebsen test was retaken with the WREX. The test times were again recorded.

Patients also were provided with a questionnaire (Table II) regarding their experience with WREX to obtain subjective input regarding viability of such a device.

III. RESULTS

For each of the Jebsen tasks, time of completion along with the number of patients who could perform the task with the WREX but could not perform the task unassisted were recorded. The results of the Jebsen test for each of the six tasks are discussed below and shown in Table III and Fig. 3. Seventeen patients completed the first round of Jebsen testing. Of these 17, four did not return to complete the second part of testing, while three others were unable to fully finish all the tasks in both rounds of testing.

Writing: This task required the patients to write a sentence on a card placed in front of them. Results showed that time increased from an average of 19 s without the WREX to 22 s with the orthosis. Eight could perform the task both with and without the WREX.

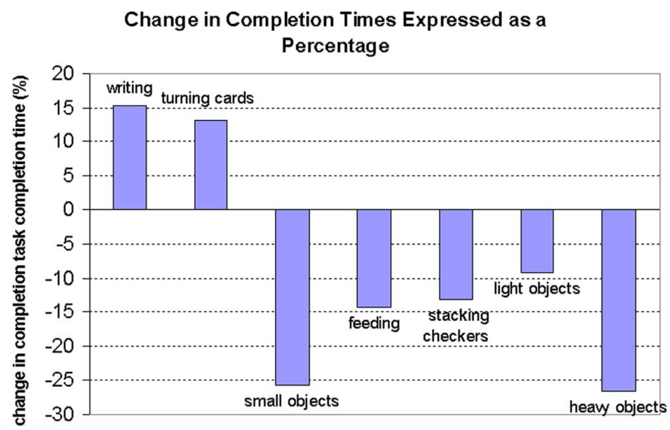

Fig. 3. Change in task completion times between performing the task without WREX and with WREX. Negative value shows an improvement in times with the WREX.

Card Turning: This test required the patients to flip over five cards lined up in front of them one at a time. Results showed that time increased from an average of 11.4 s without the WREX to 12.9 s with the orthosis. Two who were unable to perform the task unassisted were able to perform the task with the WREX.

Small objects: For this task, small common objects were picked up and dropped in a can. The average time was 21.8 s without the WREX and 16.2 s with the WREX attached. This represented a 26% decrease in time. Four patients who could not perform the task unassisted could do so with the WREX.

Feeding: This was a simulated feeding task. Average time was 28 s before the WREX and 24 s with the WREX, which represented a 14% decrease. Four who could not complete this task previously could perform the test with the WREX.

Stacking checkers: This test involved stacking four checker pieces in front of the patient. Ten patients participated. Average time was 13 s before the WREX and 11.3 s with the WREX. This did not show a significant difference between the two times.

Large light objects: This task involved moving five empty cans individually. Eight patients participated, and average time decreased from 11.9 to 10.8 s. Again, there was no significant difference in times.

Large heavy objects: This task is the same as the one above except the cans were full. Average time decreased from 21.1 to 15.5 s.

Four of the patients who took the Jebsen test without the WREX did not do the Jebsen test with the WREX. One patient stated that there was no one to attach and remove the WREX from the chair and therefore was unable to use it, even though he was a good candidate. Another patient did not use the WREX sufficiently because of excessive contractures at the elbow and shoulder. It also interfered with access to her wheelchair joystick. One patient was sufficiently functional without the WREX and it interfered with his wheelchair joystick. He also had significant contractures. The fourth patient did not use the WREX often for various reasons, including interference with teeth brushing, inability to hold utensils correctly, and difficulty with writing.

Eleven patients provided feedback for the questionnaire. The following activities were cited as most useful with WREX: eating, stretching, increasing ROM, and raising one's hand. Writing was one activity, however, that patients felt the WREX made difficult to perform. Additionally, interference with the wheelchair and tray was cited as a problem area. Some could not comfortably use their wheelchair joystick. The stationary shoulder mount restricted the ability of some to move around freely. Some also felt that their wrist motion was restricted by WREX. A suggested improvement was adding a WREX to the other hand and making it mobile so it could be attached to a table or the bedside.

There was a range of responses on the questionnaire regarding amount of use. Most participants said they used the WREX for 2–6 h a day. A few patients used it for a few minutes daily, and others did not use it much at all. All study participants responded favorably to the aesthetics of the WREX. All but one said they would like to obtain a WREX. All but one said that the device did not tire them.

Each individual was asked to comment on the orthosis subsequent to participating in the study. Most of the users exclaimed how beneficial the orthosis was in aiding with feeding as well as raising their hands in school. Many activities of daily living that were difficult to perform without the orthosis were easier to do while wearing the WREX. Some of these activities included picking up items such as forks, pens, hairbrushes, and/or toothbrushes, and playing swords and drums, as well as typing on the computer. Several patients noted that self-esteem increased while using the WREX.

A. *Case Narratives*

1) Patient 1: Patient 1 had SMA and bicep strength of 3 to 3−. Prior to using the WREX, the patient needed his left hand to assist the right hand to complete each of the tasks in the Jebsen test. After taking the WREX home and becoming familiar with it, he retook the Jebsen test. Although the patient's times in the Jebsen test increased for most of the tasks, he was now able to perform all but one of the tasks without any help from his left hand. His elevation improved as well as his reaching ability. The patient reported that the orthosis enhanced his ability to play the drums by doubling the amount of time he was able to practice.

2) Patient 2: Patient 2 had CMD, deltoid strength of 3, and bicep strength of 4. The patient reported that the orthosis helped increase the movement in her upper extremities, allowing her to accomplish tasks that she previously could not do. She had a weak grip, which made performing tasks 6 and 7 of the Jebsen test challenging. However, she was quite functional without the WREX.

3) Patient 3: Patient 3 had DMD, normal grip strength, grade 3 deltoid strength, and grade 4 bicep strength. The patient was pleased with the orthosis and claimed that it helped improve his performance in many different tasks. His total Jebsen test time decreased significantly while wearing the WREX.

4) Patient 4: Patient 4 had SMA, grip strength of 4, trace deltoid strength, and 3− strength in his bicep muscle. He scored much better on the Jebsen test while wearing the orthosis, reporting that it improved ROM and assisted with tasks such as eating spaghetti, playing chess, and writing. He would also like to have a 'sports' WREX to be able to throw a baseball. He very much wanted to keep the WREX. He continues to use the WREX daily.

5) Patient 5: Patient 5 had DMD, normal grip strength, grade 3− strength in his deltoid, and grade 3 to 3− strength in his bicep. He decreased his Jebsen test times substantially while wearing the WREX, particularly in the elevation tasks. He said that several activities, such as handraising, were easier to complete. He asked to take home one of the research prototypes at the conclusion of the study, however he was somewhat concerned about the appearance.

6) Patient 6: Patient 6 had DMD, grip strength of 4, deltoid strength of 2+, and bicep strength of 2+ to 3−. He did not use the WREX much at home because he claimed that it got hung up at the shoulder. This was due to one of the small links at the shoulder getting stuck momentarily in an awkward position. Nonetheless, he scored much better on simulated feeding and small object manipulation tasks of the Jebsen test. The patient was not very enthusiastic about using the orthosis, claiming that it restrained his arm.

7) Patient 7: Patient 7 had DMD, grip strength of 4, and deltoid strength of 3+ to 4−. The patient's times in the Jebsen test increased slightly with the WREX. Nevertheless, the patient claimed that WREX made it easier to write and raise his hand.

8) Patient 8: Patient 8 had DMD, grip strength of 4, and deltoid strength of 3. The total time of the Jebsen test decreased significantly while wearing the orthosis, particularly in the elevation tasks of simulated feeding and small-object manipulation. This patient found the WREX to be very useful in feeding and noticed that his arm felt looser after using it. His mother continually encouraged him to use the WREX.

9) Patient 9: Patient 9 had DMD, grip strength of 3, trace deltoid strength, and bicep strength of 3−. While wearing the orthosis, the patient decreased his times for several tasks including lifting large light objects, simulated feeding, and small-object manipulation. The patient reported that the orthosis aided in picking up items such as pens, forks, and paper.

10) Patient 10: Patient 10 had DMD, trace deltoid and bicep strength, and grip strength of 4. With the WREX, the patient was able to complete three of the tasks that he could not perform without the orthosis. His times in each of the other tasks decreased while wearing the WREX. He stated that the orthosis gave him a greater ROM, as well as more independence. He was able to perform a variety of tasks, which enhanced his self-esteem. It was noticed that the patient had a greater enthusiasm for all tasks when he used the orthosis. He continues to use the WREX daily at home and at school for about 5 h/day. He liked the way it looked and claimed it did not tire him. He would like to have a motorized WREX to be able to pick up heavy objects.

11) Patient 11: Patient 11 was diagnosed with a congenital myopathy with retinitis pigmentosa and ataxia and had grade 3+ strength throughout. He was too strong for the WREX. For half the tasks, the times decreased while wearing the WREX, while the others increased. He is a very active child and compensates well for his weakness. The WREX appeared to hinder his movements. As a result, he did not use it much.

12) Patient 12: Patient 12 had CMD, grip strength of 2, a deltoid strength of 3−, and bicep strength of 4−. He was fed through a tube, which significantly minimized the usefulness of the WREX. The orthosis increased his ability to raise his hand, but he claimed that the WREX prevented him from operating his wheelchair joystick. He moves between his wheelchair, his bed, and a chair quite frequently, and it was difficult to use the WREX because it was always attached to his wheelchair.

13) Patient 13: Patient 13 was the youngest and smallest participant at 13 kg. He had SMA, deltoid strength 3, and bicep strength of 3. He reported that he had more arm movement while wearing the WREX and liked it overall. He showed marginal improvement with the WREX.

14) Patients 14–17: These patients did not return for full followup.

IV. Discussion

This project tested the WREX with 17 children in a home setting. The results obtained were generally positive; most of the children found the WREX beneficial. Four children found it to be indispensable and continue to use the prototypes. One subject has purchased a WREX for his other arm as well. Subjects have come to rely on the WREX for activities such as eating, pick-and-place activities, and exercise. The WREX also allows the children to independently participate in group activities such as painting in class or eating a meal with their family.

The children who found it most beneficial seemed to be the midrange disabled children, particularly those with spinal muscular atrophy. The weaker children often did not have enough strength to overcome the inertia of WREX. The stronger children tended to stick with their compensatory movements techniques to overcome limitations rather than have an additional device. Strong parental support was a big factor in successful use of WREX, particularly in the early trial phase. The novelty factor of the device was not really apparent in these trials, or if it was a factor, it became apparent within the first visit if this subject would use the WREX. If a potential subject did not fall within the selection criteria despite being enthusiastic about a "robotic" device, they were not selected. This was likely due to them being either too weak, too strong, or having severe contractures.

Although liked by most users, there were a few who did not see a benefit of the extra "gadget" attached to them. These patients relied on compensatory movements or their caregiver to perform their movements. They found it difficult to adjust to something new. One patient in particular clearly benefited from the WREX and scored much better on the Jebsen test yet refused to use the WREX because it got "hung up" at the shoulder. This was easily fixed with a minor adjustment, but he remained apprehensive. Some of the children were quite intolerant of the WREX's interference with their environment, whether it was their armrest, joystick, or clothes. To be acceptable, any permanent assistive device attached to the human as intimately as the WREX must provide uncompromising functionality. Users were quite forgiving of the aesthetics of the WREX. Two colors were used, blue and silver. The blue color was suggested by two of the users. We, therefore, sandblasted the metal parts and spray-painted them a light blue. This gave the device a pleasing, finished appearance.

The Jebsen test was chosen as the most suitable metric for everyday tasks despite it being designed to evaluate *both* hand and arm function, whereas the primary support of the WREX is in arm function. This did result in some of the Jebsen tasks, such as writing and stacking checkers, providing minimal indication of efficacy of the device. An alternative test, the Wolf Motor Function Test [22], was relatively new at the time of testing and, therefore, not used. This may, however, be a more appropriate test for arm function despite having a number of tasks that address finger dexterity.

Tasks 3 and 4 of the Jebsen were small-object manipulation and simulated feeding, which highlighted the utility of the WREX. In these tasks, elevation of the elbow was required. This was not possible for most of the users before the WREX. With the WREX, however, everyone who retook the Jebsen was able to perform these tasks. The disadvantage of the Jebsen test, as mentioned earlier, was that for some of the tasks finger and wrist dexterity were required, which was not always present. The WREX was not designed to assist in this area of fine motor control. The overall Jebsen test scores were not provided, as is customary, because few subjects completed all tasks with and without the WREX. Rather, times are presented for individual tests such as small object manipulation and simulated feeding which highlight the effect of WREX.

The number of patients in the study was too small to achieve statistical significance, particularly with the multiple disabilities and high variability within each condition. The results, nevertheless, demonstrated a positive trend and showed the WREX had a positive impact on most of the children tested. We should note that adjusting the WREX to each individual (e.g., adjusting the link lengths and the number of bands, and precisely positioning and orienting the shoulder pivot) required much skill. The patients tested in the early part of the protocol may have been at the bottom of this "learning curve" for the research team. Individualized adjustments were also made, such as placing an extra rubber band at the elbow joint to assist in elbow flexion. This was done for two of the weaker patients who had difficulty reaching their mouths. The WREX has undergone modifications

Fig. 4. New JAECO WREX.

since the testing and has been made easier to attach to a wheelchair and customize to individual users. This is a result of experience gained from the testing phase.

Task 1 of the Jebsen test, writing, was actually worse with the WREX because the hands hovered over the table, which made precise control of writing difficult. Task 2, flipping cards over, required the ability to pronate/supinate, which the WREX did not provide. Other tasks such as the simulated feeding also required a good degree of supination/pronation, which a lot of the subjects did not have. Therefore, some of the times suffered due to the absence of this movement. A supination/pronation provision is currently being investigated for the WREX.

The WREX recently has been licensed to JAECO Orthopedics Inc., Hot Springs, AR, and is available for sale. The latest version of WREX is shown in Fig. 4. The use of WREX as an upper-extremity therapeutic tool is also currently being investigated with the potential of affecting a much larger population such as people who have had a stroke. Work also continues in the area of using motors as a power-assist adjunct to the WREX. This would enable picking up heavier objects and facilitating movement for profoundly weak subjects.

V. Conclusion

Due to decreasing motor strength, children and adults with neuromuscular diseases frequently come to rely on others for assistance in activities of daily living. This study has developed a mechanical assistive device that restores some function to these children. Most patients in our study demonstrated both quantitative and qualitative benefits from wearing the WREX. This device increased upper extremity ROM, decreased the amount of time required for activities of daily living, enhanced overall performance, and raised self-esteem. A number of children continue using the WREX daily.

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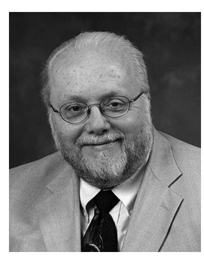

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