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Universal Exercise Unit for Treatment of a Child Following Hemispherectomy: A Case Report

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Universal Exercise Unit for Treatment of a Child Following Hemispherectomy: A Case Report

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Abstract

Background: Rasmussen's encephalitis is a rare neurological disorder, known for unilateral inflammation of the cerebral cortex, most commonly occurring in children.¹ The only known cure for the seizures associated with the syndrome is cerebral hemispherectomy. Physical therapy following this procedure is vital for return to prior level of function and to optimize independence. One rehabilitation tool is The Universal Exercise Unit (UEU), a form of body weight supported training that uses a system of bungee cords and pulleys to facilitate or resist movements. The purpose of this case is to describe the Universal Exercise Unit (UEU) as an intervention for a child following hemispherectomy surgery for Rasmussen's encephalitis. **Case Description:** A nine-year-old girl underwent cerebral hemispherectomy about 1 year prior to rehabilitation in the Universal Exercise Unit. **Intervention:** The patient completed two 8-week intensive outpatient sessions approximately 6 months apart, consisting of three 60 minute sessions per week inside the Universal Exercise Unit (UEU). **Outcome Assessment:** The "Universal Exercise Unit Skills Assessment" was the primary outcome assessment used, consisting of 18 functional items, scored 0-6 based on level of assistance needed to complete each task. **Discussion:** This case describes a unique rehabilitation treatment option for patients who have undergone cerebral hemispherectomy. The Universal Exercise Unit Skills Assessment scores for the patient in this case study improved in both 8-week intensive sessions, and the patient demonstrated functional improvements in kneeling and half kneeling, half kneel to stand transfers, sit to stand transfers, and single leg stance. These improvements suggest that the Universal Exercise Unit may be a beneficial intervention for children following hemispherectomy.

Background

Rasmussen's encephalitis is a rare neurological disorder, known for unilateral inflammation of the cerebral cortex, most commonly occurring in children under the age of 10.¹ The syndrome was first described by Rasmussen et al. in 1957, who examined three patients who had chronic focal seizures and damage to one of the cerebral hemispheres.²

Symptoms that commonly are associated with the syndrome include severe and frequent seizures, loss of motor skills and speech, hemiparesis, brain inflammation, and mental deterioration.¹ Rasmussen's encephalitis seems to act as an autoimmune disease where immune cells enter the brain and cause inflammation and damage, drug-resistant epilepsy, and progressive neurocognitive deterioration.¹ Some studies show varied success of treatment with corticosteroids, intravenous immunoglobulin, or tacrolimus, all of which suppress or modulate the immune system.¹ However, cerebral hemispherectomy remains the only cure for the seizures associated with Rasmussen's encephalitis.³

The Universal Exercise Unit was developed as a part of the Thera Suit method. It is a system of pulleys, straps, and harnesses attached to a metal cage to perform various exercises to improve functional abilities (see Figure 1). According to Thera Suit, there are two main uses of the Universal Exercise Unit.⁴ The first is a system of pulleys and that are utilized while the patient is treated on a bed or chair. In this use of the UEU, the goal is to improve strength, range of motion, and flexibility.⁴ The UEU eliminates the effects of gravity by supporting the patient's body weight, and therefore allows the patient to initiate movements regardless of strength. The therapist can add various weights to the pulleys to assist or resist movements, thereby making them easier or more challenging based on the patient's functional level. The main goals of this application, according to Thera Suit, are to prevent atrophy, increase strength, increase range of motion, improve flexibility, and prevent joint contractures.⁴

The second way the UEU is utilized is commonly referred to as the "spider cage."⁴ In this application, children wear a harness belt with 8-10 bungee cords attached from the belt to the cage. The cords can be attached to the cage higher than the belt to unload weight and to aid with certain tasks, or below the belt to increase resistance thereby making activities more challenging. When the cords are attached higher than the patient's belt, the UEU acts as a means of providing body weight supported training for various positions, such as quadruped, tall kneeling, or standing. The benefit of this system is that the therapist has free hands to aid in the movement, while still allowing the patient to demonstrate his or her independence in a setting that he or she might not otherwise be able to do.

While the UEU is commercially available, not all therapists may be familiar with its application outside of more standard diagnoses, such as CP. Thus, the purpose of this case study is to describe the use of the Universal Exercise Unit as treatment for a child following hemispherectomy for seizures associated with Rasmussen's encephalitis.

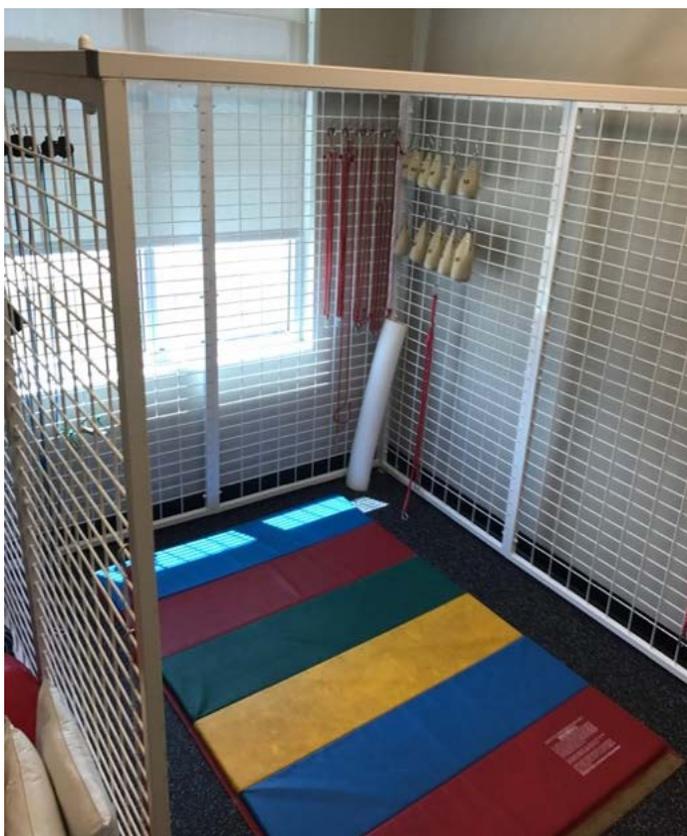


Figure 1- The Universal Exercise Unit (UEU).

Case Description

The patient was a 9-year-old girl with a history of right-sided hemispherectomy about 1 year prior to intervention in the UEU for outpatient physical therapy. She had a history of epilepsy partialis continua and Rasmussen encephalitis which affected all activities of daily living. Prior to surgical hemispherectomy, the young girl was wheelchair-bound because she had difficulty walking, and she was mostly dependent on caregivers for daily tasks. Her symptoms began approximately 7 months prior to surgery with left-sided shaking and then progressed to continuous jerking and episodes with left eye deviation. She and her family sought care from neurology physicians, who continued her antiepileptic medications and administered intravenous immunoglobulin. Her seizures became stable, but very consistent, so an MRI and PET scan were performed and they demonstrated a low area in the right posterior dorsal medial frontal lobe and an area of asymmetrically increased uptake in the right frontal lobe. As treatment for Rasmussen syndrome, the patient underwent a hemispherectomy of the right cerebrum. Her hospital course was complicated by a bone infection for which a washout was performed as well as a craniotomy with bone flap removal. At the time of the patient's outpatient evaluation two months after surgery, her current medications included Keppra and gabapentin.

The patient was referred to the pediatric outpatient clinic after being discharged from a transitional care unit where she had inpatient care for approximately two months. A full physical therapy evaluation was completed, including observation of her ability to complete functional tasks in all positions. In sitting, she tended to demonstrate left trunk lean with left shoulder depression, but could actively correct to midline with verbal cues. In stance, left knee hyperextension was present. When ambulating, the physical therapist noted a significant left hemiparetic gait with left knee hyperextension during the stance phase. She demonstrated left lower extremity weakness during functional gross motor activities, which placed her at a high risk for falling. She also was unable to complete age-appropriate gross motor skills such as jumping, skipping, and hopping. She wore a hinged AFO on her left lower extremity secondary to foot drop that occurred as a result of the surgical intervention.

The patient had received about 1 year of conventional physical therapy consisting of 1 session per week for 30-45 minutes per session prior to the interventions described in this case. She had demonstrated increased independence with walking and navigating stairs, but still had difficulty with sequencing involved with skipping and jumping. She had an overall optimistic outlook on her condition and had a personality consistent with a 9-year-old girl.

Intervention

The patient participated in 2 periods of intensive therapy sessions, each consisting of one hour of physical therapy in the Universal Exercise Unit, three times per week for eight weeks, separated by approximately 6 months. She was selected to receive this type of therapy after completing about 1 year of conventional physical therapy once per week after her surgery. She could walk without an assistive device and navigate stairs, but still lacked in activities that required isolation of her left lower extremity, specifically single leg stance activities. Much of her movement required assistance of her right lower extremity to compensate for left lower extremity weakness.

The patient, her mother, and the physical therapist developed quarterly goals to progress with developmental skills. The patient's goals at the time of the first Universal Exercise Unit intensive session included: left single leg heel raises with minimal upper extremity support for balance, left single leg stance while performing multidirectional taps with right leg, improved strength in major hip muscle groups by 1 manual muscle test grade, improved left knee strength by 1 manual muscle test grade, and for the patient and family to be independent in their home exercise program.

While the patient was suspended in the UEU, the physical therapists employed several strategies to improve function. Much of the patient's interventions involved reaching with each hand outside her base of support to the left to encourage weightbearing onto her left leg. She was challenged to perform single leg stance on her left leg while completing dynamic upper extremity tasks such as

puzzles and playing catch. Another method used to encourage single leg stance was kicking a ball with her right leg, forcing weightbearing onto the left leg. Additionally, the suspension provided from the UEU gave the patient the necessary support and balance to be able to complete both double leg and single leg jumping from a squatting position. She was also challenged to perform half kneel to stand transfers bilaterally and sit to stands from various surface heights. Occasionally, standing tasks were performed on a wobble board to ensure equal bilateral weightbearing and to challenge the patient's balance.

Outcomes Assessment

Because there is no formalized evaluation tool for evaluating the effectiveness of the Universal Exercise Unit as a treatment, a physical therapist at the outpatient clinic developed a tool, referred to as the Universal Exercise Unit Skills Assessment. This tool evaluated 18 functional tasks when the child was outside of the UEU: sitting, quadruped, quadruped with reaching, short kneel, tall kneel, half kneel (left and right), half kneel to stand (left and right), sit to stand, standing, weight shifting, single leg stance (left and right), jumping, single leg jump (left and right), and marching. Each task was rated based on level of independence using a 0 (independent) to 6 (maximum assistance of 2 or more people) scoring rubric. The sum of the item scores, ranging from 0 to 108, is used as the global assessment score, where lower scores represent increased independence and higher scores represent poorer function. In addition, most tasks were given a percentage of accuracy at the therapist's discretion or a description of important observational information, as seen in Tables 1 and 2.

The patient described in the above case performed two intensive therapy sessions, consisting of one hour of physical therapy in the Universal Exercise Unit, three times per week for 8 weeks, on two different occasions separated by approximately 6 months. At her first intensive session, the patient's pre-UEU assessment score was 22. Areas of deficit included quadruped with reaching, short and tall kneeling, bilateral half kneeling, bilateral half kneel to stand transfers, sit to stand, bilateral single leg stance, and bilateral single leg jumping (Table 1). After her 8-week intensive session, the patient's ability improved to a score of 8. She demonstrated improvement in all areas of deficit except for single leg jumping, where she continued to require at least moderate assistance on her left lower extremity. Most importantly for application to daily activity, the patient's mother reported functional improvement in standing endurance and navigating stairs, suggesting a degree of construct validity for the assessment.

The patient's second 8-week intensive session began about 6 months after the first session terminated. At that time, the Universal Exercise Unit Skills Assessment was re-evaluated. The patient's initial score in the UEU was a 10, with the only skills requiring any form of assistance were half kneel to stand bilaterally and single leg jumping. Following the second 8-week intensive physical therapy session, the patient's final score was 3, with her only limitation being single leg jumping on her left lower extremity (see Table 2)

Table 1- Universal Exercise Unit Skills Assessment scores and descriptions for the patient’s first intensive UEU session.

| Task | Initial Score | Description | Final Score | % accuracy |
|---|----------------------|---------------------------------------|--------------------|---|
| Sitting | 0 | 100% accurate | 0 | 100% accurate |
| Quadruped | 0 | 90% accurate | 0 | 100% accurate |
| Quadruped with reaching | 1 | 70% accurate | 0 | 90% accurate |
| Short kneel | 1 | 95% accurate (leans to right) | 0 | 100% accurate |
| Tall kneel | 1 | 95% accurate (leans to right) | 0 | 100% accurate |
| Half kneel (left) | 2 | 50% accurate | 0 | 95% accurate |
| Half kneel (right) | 2 | 50% accurate | 0 | 95% accurate |
| Half kneel→stand (left) | 3 | 70% accurate | 1 | 90% accurate |
| Half kneel→stand (right) | 4 | 70% accurate | 2 | 80% accurate |
| Sit to stand | 1 | 70% accurate | 0 | 90% accurate- more weight on right (70%) |
| Standing | 0 | | 0 | |
| Weight shifting | 0 | Full to right, 70% accurate to left | 0 | Full bilaterally but knee lock out on right |
| Single leg stance (left) | 2 | 2 seconds | 0 | 2 seconds |
| Single leg stance (right) | 0 | 15 seconds | 0 | 20 seconds |
| Jumping | 0 | 60% accurate (all weight on right LE) | 0 | 80% accurate, most weight on right |
| Single leg jump (left) | 4 | 1 jump | 4 | 1 jump not leaving ground |
| Single leg jump (right) | 1 | 5 jumps | 1 | 5 jumps |
| Marching | 0 | | 0 | 95% accurate |
| Global Assessment Score | 22 | | 8 | |
| 0-Independent 3-Minimal assistance 1-Standby assistance 4-Moderate assistance 2-Contact guard assistance 5-Maximum assistance 6-Maximum assistance of at least 2 people | | | | |

Table 2- Universal Exercise Unit Skills Assessment scores and descriptions for the patient’s second intensive UEU session.

| Task | Initial Score | % accuracy | Final Score | % accuracy |
|---|----------------------|---|--------------------|---------------------------|
| Sitting | 0 | 100% accurate | 0 | 100% accurate |
| Quadruped | 0 | 90% accurate | 0 | 90% accurate |
| Quadruped with reaching | 0 | 70% accurate | 0 | 80% accurate |
| Short kneel | 0 | 90% accurate | 0 | 90% accurate |
| Tall kneel | 0 | 100% accurate | 0 | 100% accurate |
| Half kneel (left) | 0 | 100% accurate | 0 | 90% accurate |
| Half kneel (right) | 0 | 100% accurate | 0 | 90% accurate |
| Half kneel→stand (left) | 3 | 80% accurate | 0 | 80% accurate |
| Half kneel→stand (right) | 3 | 80% accurate | 0 | 90% accurate |
| Sit to stand | 0 | 70% accurate (leans to right) | 0 | 100% accurate |
| Standing | 0 | 95% accurate | 0 | 100% accurate |
| Weight shifting | 0 | Full bilaterally, but knee locks out on right | 0 | 100% accurate |
| Single leg stance (left) | 0 | 1-2 seconds, 90% accurate | 0 | 1-2 seconds, 90% accurate |
| Single leg stance (right) | 0 | 10 seconds, 100% accurate | 0 | 17 seconds, 100% accurate |
| Jumping | 0 | 90% accurate | 0 | 90% accurate |
| Single leg jump (left) | 4 | Feet not leaving ground | 3 | Feet not leaving ground |
| Single leg jump (right) | 0 | | 0 | |
| Marching | 0 | 95% | 0 | 100% accurate |
| Global Assessment Score | 10 | | 3 | |
| 0-Independent 1-Standby assistance 2-Contact guard assistance 3-Minimal assistance 4-Moderate assistance 5-Maximum assistance 6-Maximum assistance of at least 2 people | | | | |

Discussion

Functional improvements noted in the case described above suggest that the UEU may be a beneficial treatment intervention utilized for patients following cerebral hemispherectomy. This case is an example of a unique treatment option being utilized for a child with a rare disease and rare surgical intervention.

There are many benefits of utilizing the Universal Exercise Unit in a child following hemispherectomy as a result of Rasmussen’s encephalitis. First, it provides a means of body weight supported training while allowing the physical therapist to be hands-free for providing assistance as needed. Second, it provides psychological benefit for the patient by allowing him or her a sense of independence that he or she may not otherwise experience. Children are able to stand, jump, or even dance in a safe, controlled setting, which in turn may improve not only their physical function, but also

their overall quality of life. The patient tolerated treatment in the UEU well and seemed to enjoy coming to her physical therapy sessions. This was likely partially due to her positive attitude about her condition, and partially due to the increased level of independence that the UEU provided her. She was able to complete tasks in the UEU that she was otherwise not able to do, and functional gains in the UEU translated to her daily life. This included improvements in sitting balance, gait pattern, and ease of riding her bicycle at home with her siblings.

There are multiple reasons that this patient may have demonstrated functional improvements once beginning her 8-week sessions in the UEU. First, the Universal Exercise Unit provided a unique treatment option for the patient and was a change in type of treatment she had received prior to the intensive session. Second, the patient was seen three times per week for hour-long sessions. This was an increase in the frequency and duration of her treatment sessions, as she was being seen only 1 time per week since her procedure was done. In a study done by Fritz et. al, 19 participants who had undergone cerebral hemispherectomy received Intensive Mobility Training, consisting of overground walking, body weight supported treadmill training, dynamic and static balance activities, range of motion and strengthening activities, and neuromuscular re-education. These sessions 3 hours per day for 10 days, and in general the participants demonstrated improvements in the Berg Balance Test, Six Minute Walk Test, and Dynamic Gait Index, among others.⁵

While the Universal Exercise Unit has not been thoroughly examined for the patient population following hemispherectomy, it has been used for treatment of other diagnoses. In a 2015 study done by Afzal et. al, the Universal Exercise Unit was examined as a treatment option for 25 children diagnosed with cerebral palsy. The outcome measure used in this study was the Gross Motor Function Measure (GMFM-88). The participants were seen 5 days per week for 1 hour treatment sessions, spanning 3 months duration. This study concluded that the Universal Exercise Unit is an effective tool when used in combination with traditional therapy for demonstrating improvement in gross motor and functional skills for children age 3 to 14 who are diagnosed with cerebral palsy.⁶

The outcome measure described in this case was specifically developed for use with the UEU by clinicians at the outpatient pediatric facility where the patient's care took place. While it is a useful tool for examining level of assistance required for a variety of functional tasks, the validity or reliability of this tool as a standard outcome measure remains as yet unknown. Thus, the use of this outcome is a potential limitation of this case study, and future research may benefit from the use of a validated, reliable outcome measure. Two possible outcome measures that may be beneficial for this diagnosis in particular are the Functional Independence Measure for Children (WeeFIM) and the Pediatric Evaluation of Disability Index (PEDI).

The Functional Independence Measure for Children (WeeFIM) is an adaptation of the Functional Independence Measure (FIM) that is commonly used in adult rehabilitation, but is specifically tailored for children. It is typically used for children from 6 months to 7 years, but may also be used for children older than 7 years if their functional ability is below those children 7 years old without functional disabilities, up to 21 years old.⁷ It was based on a framework of pathology, impairment, disability and handicap, and the "burden of care", as described by the World Health Organization in 1980.⁶ This tool is an 18-item instrument that measures a child's performance in daily activities in the domains of self-care, mobility, and cognition. Each of the items is rated on a 1-7 scale where 1 indicates total assistance and 7 indicates independence. For each task, there is no concern about the safety or amount of time required to complete the task, but unsafe technique or increased amount of time to complete is scored as modified independence (6). Tasks specific to mobility tasks in physical therapy in this assessment tool include: bed to chair transfer, toilet transfer, tub/shower transfer, walking, and stairs. A pilot study done by Msall et al. demonstrates that the WeeFIM is a valid measure for tracking disability in preschool and young children with limb deficiency, Down's syndrome, spina bifida, cerebral palsy, and extreme prematurity.⁸

The Pediatric Evaluation of Disability Inventory Computer Adaptive Test (PEDI-CAT) is a judgment-based measure that requires nothing more than a testing software to complete and the child's parent or primary caregiver answers questions based on the child's functional performance at the

current time.⁹ This tool is designed to be used for children aged birth through 20 with physical and/or behavioral conditions.⁹ The PEDI-CAT measures abilities in the 3 functional areas of Daily Activities, Mobility, and Social/Cognitive, and a fourth domain, the Responsibility domain, reports whether the child or caregiver takes responsibility for multifaceted daily tasks.¹⁰ There are 75 mobility items in four areas that are most relevant to physical therapy: Basic Movement and Transfers, Standing and Walking, Steps and Inclines, and Running and Playing. All respondents begin with the same item that falls in the middle of the range of difficulty, and the response to that item dictates which item will appear next, whether it's easier or harder. This makes the PEDI-CAT a very custom tool with few irrelevant items to the child. Once the predetermined stopping rule has been satisfied, the test is completed, and scores are displayed immediately.¹⁰ Validity of the PEDI-CAT was established through a review of existing adult and pediatric functional assessments and input from practicing clinicians, parents of children with and without disabilities, and experts in the field of rehabilitation.⁹ In a 2012 study, the PEDI-CAT was able to differentiate between groups of children with and without disabilities based on parent responses and the scores of the PEDI-CAT Mobility domain were shown to differentiate the functional mobility status between groups of children who used an assistive device (higher scores) or wheelchair (lower scores).⁹ Test-retest reliability results also have been reported as high (0.96–0.99) for all 4 domains when parents completed the PEDI-CAT twice within 1 month's duration.⁹

In conclusion, this case highlights the functional improvement noted in a single patient with Rasmussen's encephalitis following the use of the UEU on two separate occasions. The UEU provided a safe and enjoyable intervention for this 9-year-old girl with a long history of therapy treatment. Other children with history of a hemispherectomy may benefit from the Universal Exercise Unit intensive sessions as part of their physical therapy treatment, but each patient should have an individualized program of functional activities (Figure 2). Future research is needed to evaluate the Universal Exercise Unit as an intervention for pediatric physical therapy for a variety of diagnoses, as well as valid outcome measures that can be used to evaluate its effectiveness in clinical settings.

Functional activities that can be performed in the UEU:

- Quadruped with/without reaching
- Short kneeling
- Tall kneeling with/without reaching for weight shift
- Half kneeling with/without reaching for weight shift
- Half kneel to stand transfers
- Standing
- Weight-shifting in standing
- Single leg stance
- Sit to stand from various heights
- Double or single leg squat
- Double or single leg jumping
- Marching

Figure 2- Exercise options for patients in the UEU.

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