

Spring 2015

Implementation of a Land and Aquatic-Based Physical Therapy Program for a 6-Year-Old Male with Aicardi-Goutieres Syndrome: A Case Report

Kristin A. Cheney
Governors State University

Follow this and additional works at: <http://opus.govst.edu/capstones>

 Part of the [Physical Therapy Commons](#)

Recommended Citation

Cheney, Kristin A., "Implementation of a Land and Aquatic-Based Physical Therapy Program for a 6-Year-Old Male with Aicardi-Goutieres Syndrome: A Case Report" (2015). *All Capstone Projects*. 109.
<http://opus.govst.edu/capstones/109>

For more information about the academic degree, extended learning, and certificate programs of Governors State University, go to http://www.govst.edu/Academics/Degree_Programs_and_Certifications/

Visit the Governors State Physical Therapy Department

This Project Summary is brought to you for free and open access by the Student Capstone Projects at OPUS Open Portal to University Scholarship. It has been accepted for inclusion in All Capstone Projects by an authorized administrator of OPUS Open Portal to University Scholarship. For more information, please contact opus@govst.edu.

**IMPLEMENTATION OF A LAND AND AQUATIC-BASED PHYSICAL
THERAPY PROGRAM FOR A 6-YEAR-OLD MALE WITH
AICARDI-GOUTIERES SYNDROME:
A CASE REPORT**

By

Kristin A. Cheney, SPT
B.S., Olivet Nazarene University, 2011

CAPSTONE PROJECT

Submitted in partial fulfillment of the requirements

For the Degree of Doctor of Physical Therapy

Governors State University
University Park, IL 60484

2015

ACKNOWLEDGEMENTS

I would like to express my gratitude to everyone who supported me throughout the course of this capstone project. I am thankful for their guidance, constructive criticism, and friendly advice during the project work.

I express my warm thanks to my clinical instructor, Dr. Wendy Reils, my faculty advisor, Dr. Roberta O'Shea, my reviewer Dr. Sarah Bugajski, as well as my classmate and future colleague Ms. Sarah Wicklin for all of their support and guidance.

ABSTRACT

Background & Purpose: There is little research surrounding Aicardi-Goutieres Syndrome (AGS) in all aspects of patient care, including physical therapy. AGS, a rare, early-onset, neurodegenerative disorder, includes symptoms of postnatal microcephaly, spasticity, muscle hypotonia, and developmental delay. The purpose of this study was to examine the effectiveness of a land and aquatic-based physical therapy treatment program for improving functional limitations and balance for a 6 year-old male referred to physical therapy for developmental delay secondary to an onset of AGS.

Case Description: This case study included a land and aquatic-based physical therapy treatment program focused on neuromuscular re-education, strengthening exercises, weight-bearing activities, and passive range of motion exercises in order to slow the patient's decline of function. To measure the impact of the intervention the Pediatric Evaluation of Disability Inventory (PEDI) was used to measure capability and performance of selected functional activities within the domains of self-care, mobility, and social function. Balance was also observed in a variety of positions in order to determine how long a patient could maintain a given position unassisted.

Outcomes: The patient demonstrated an increase in his ability to sit unsupported in a tailored-sitting position from one second to five seconds. The patient also demonstrated a slight decrease in a few scores on the PEDI, including the Functional Skills domains of Mobility (20.9 to 18.2), Social Function (39.6 to 37.9), and the Caregiver Assistance domain of Social Function (20.4 to 11.3).

Discussion: The results from this case report demonstrated that a physical therapy treatment program may positively impact functional balance in a child with AGS, however it may not contribute to improvements in the patient's ability to perform overall functional activities. Additional research could include longitudinal studies starting immediately post diagnosis of AGS, as children may live with the affects of AGS for several years post diagnosis.

BACKGROUND/ PURPOSE

Aicardi-Goutieres syndrome (AGS) is a rare, early-onset, neurodegenerative disorder first discovered in 1984. AGS is primarily autosomal recessive with onset typically within the first four months of age.¹ The main symptoms of AGS include postnatal microcephaly, spasticity, abnormal eye movements, muscle hypotonia, vomiting and feeding difficulties, irritability, developmental delay, and loss of any psychomotor skills a child may have acquired prior to onset. It is not uncommon for children affected by AGS to also have a sibling with the disorder as well, as there is a strong familial component.^{1,2}

Although most children have an early onset of AGS within the first four months of life, some children show later onset around 7 and 12 months of age. Due to the very early onset of this disorder and its rapid progressiveness, children often do not make significant gains in psychomotor skills. The cognitive status of those affected often declines rapidly after the onset of AGS, leading to a vegetative state within childhood.³

AGS is difficult to medically diagnose, not only due to the rarity of the disorder, but also because symptoms of AGS mimic other disorders, such as cerebral palsy. Diagnosis is made based on a child's symptoms, brain abnormalities seen through imaging, and through testing of cerebrospinal fluid for a specific immune system cell and a substance known as interferon-gamma. The prognosis of a child affected by AGS depends on the severity of

symptoms, and children with an earlier onset of the disorder have a higher risk of death than those with a later onset.⁴ Although the severity of AGS varies widely depending on its time of onset, around 25% of affected children die between the age of 1 and 17 years of age.³

There is little research surrounding AGS in all aspects of patient care, including physical therapy.¹ This case report is needed to broaden the world's understanding of how a child with AGS may present and the outcomes a physical therapy program may have for a child with AGS.

The purpose of this study was to examine the effectiveness of a land and aquatic-based physical therapy treatment program for improving functional limitations and balance for a 6 year-old male referred to physical therapy for developmental delay secondary to an onset of AGS. The treatments used in this study focused on neuromuscular re-education, strengthening exercises, weight-bearing activities, and passive range of motion (PROM) exercises in order to slow the patient's inevitable decline of function and improve the patient's quality of life.

CASE DESCRIPTION

Patient History

The patient for this case study was a 6-year-old Caucasian male who presented with a recent diagnosis of the genetic disorder AGS. The patient's sister, age 9, also had been recently diagnosed with the same genetic

disorder. The patient was initially referred to physical therapy at 15 months old, secondary to developmental delay. Upon initial examination at that time, the patient presented with low muscle tone, weakness, atypical movement patterns, and significant delays in meeting developmental milestones.

The patient was followed by pediatric medicine (for routine illnesses and check-ups), pediatric neurology, genetics, and orthopedics. The patient had not been hospitalized for any trauma or illness. The patient had no known allergies, was taking no medication, and had no history of surgeries or seizures. The patient's hearing, vision, and imaging studies all demonstrated normal results. The patient had been undergoing genetic testing for several years with negative results for any disorder, and so he was presumed to have a diagnosis of cerebral palsy spastic quadriplegia. AGS presents somewhat similarly to that of cerebral palsy spastic quadriplegia. In early 2014, the patient and his sister were diagnosed with AGS through genetic testing. In addition to physical therapy, the patient also received occupational therapy and speech therapy.

Upon the patient's physical therapy re-evaluation in the summer of 2014, the start of this case report's episode of care, the patient presented as being generally content, with only occasional demonstration of irritability, and he tolerated handling by others well. Typically, the patient had low

arousal status and low affect but would occasionally respond to verbal stimuli with a smile or coo.

The patient exhibited variable needed assistance in all positions. In general, the patient required maximum assistance to maintain any position beyond supine, prone, or sitting. The amount of assistance needed on a given day depended on the amount of fatigue the patient was experiencing, what activities the patient participated in that day, and also the presence of his caregivers during therapy. It was a goal of therapy to decrease the amount of assistance the patient needed for maintaining different positions. The patient also required being carried or pushed in a wheelchair in order to maneuver around his environment. The patient demonstrated an influence of primitive reflexes and had difficulty with midline activities secondary to influence of a dominant asymmetrical tonic neck reflex (ATNR) that affected his bilateral upper and lower extremities. The patient's ability to use his arms was linked to his head position secondary to the influence of his symmetrical tonic neck reflex (STNR). The patient also demonstrated a startle reflex that occurred secondary to sudden noises or movements.

The patient required the use of bilateral solid ankle-foot orthoses to maintain proper skeletal alignment when participating in supported standing activities, as the patient demonstrated spasticity through his lower extremities secondary to the influences of primitive reflexes. The patient used a manual wheelchair with a rotational seating system. At home, the

patient used a stander and a high-low chair and was cared for by his grandparents, his legal guardians. The patient's caregivers had long-term goals to increase his independence in activities of daily living, such as feeding, dressing, and play.

The patient was chosen for study in this case report in order to widen the knowledge base on AGS in the field of physical therapy, as there is currently very little research on the syndrome and what interventions are typically employed for physical therapy treatment.

Systems Review

Neuromuscular findings

Patient presented with severe hypotonicity throughout his entire trunk and extremities. The patient also demonstrated decreased head control as a result of the hypotonicity, decreased sitting and standing balance with impaired balance strategies, decreased communication skills, and dominant influence of ATNR and STNR. The influence of ATNR and STNR resulted in spasticity throughout the patient's bilateral upper and lower extremities when the patient was placed in certain positions. The patient also demonstrated a startle reflex that occurred secondary to sudden noises or movements.

Musculoskeletal findings

Patient presented with decreased overall strength in all four extremities and neck, as well as decreased passive range of movement for all joints secondary to spasticity.

Cardiovascular findings

Patient demonstrated typical findings with regards to his cardiovascular system.

Integumentary findings

Patient demonstrated typical findings with regards to his integumentary system.

CLINICAL IMPRESSION #1

The patient presented with significantly decreased motor skills and independence in daily activities due to influence of primitive reflexes, decreased strength and passive range of motion, and other deficits consistent with the diagnosis with AGS. Due to the progressive nature of the disorder, the goal of treatment was to slow the patient's inevitable decline of function through the course of a land and aquatic-based physical therapy program.¹

The Pediatric Evaluation of Disability Inventory (PEDI) was chosen as a test and measure to track changes over a small amount of time. The PEDI is sensitive enough to show even the smallest amount of change in a short

amount of time. This is important as AGS is progressive and the patient may show minimal, if any, progress over time. The PEDI also measures the amount of caregiver assistance for several daily activities which made the PEDI an appropriate measure since one of the patient's caregivers' goals was to increase the patient's independence.⁵

The patient's ability to balance in tailored-sitting, quadruped, tall kneeling, half kneeling, and standing was observed and documented as well, as it was a short-term goal to increase the patient's time spent in tailor-sitting unassisted so the patient could observe and interact with his environment. Treatments focused on decreasing the amount of assistance the patient needed in maintaining these balance positions, as the patient would demonstrate variable assistance depending on the day. Balance observation was chosen as an important measure because, according to a study by Maria Ragnarsdottir in the journal *Physiotherapy*, balance is "vital to the performance of activities of daily living" and good balance control is "the foundation of independent mobility."⁶

EXAMINATION

Tests and Measures

Pediatric Evaluation of Disability Inventory (PEDI)

The PEDI was implemented at the start and end of the patient's episode of care. It was chosen because it provides an evaluation of

functional capabilities, performance, and changes in functional skills in children with disabilities. The PEDI can be completed by the patient's caregiver or in the form of a structured interview and "measures capability and performance of selected functional activities within the domains of self-care, mobility, and social function on three scales: Functional skills, Caregiver assistance, and Modifications." The PEDI is also very functional because it focuses on things that may not be seen in the clinical setting, such as activities observed at home.⁵

According to a study by Berg *et al.* the PEDI had an inter-rater reliability from 0.95-0.99, an inter-respondent reliability from 0.64-0.74, and an intra-rater reliability from 0.95-0.99.⁵ These numbers suggest that the PEDI has good to excellent overall reliability. According to another study by Feldman *et. al* concurrent validity of the PEDI was supported by moderately high correlations between the Battelle Developmental Inventory Screening Test (BDIST) and the PEDI summary scores.⁷ The BDIST is a "standardized assessment with developmental and adaptive content" and was chosen as a comparison to the PEDI because it assesses "similar content areas" such as dressing, toileting, and mobility."⁷ This study also supported construct validity of the PEDI, as demonstrated by significant differences between the disabled and nondisabled groups' PEDI scores and by discriminant analysis identifying PEDI scores as better group discriminators than the BDIST scores.⁷

For each domain of the PEDI three scales are calculated including functional skills, caregiver assistance, and modifications. Total scores are calculated by summing items within each domain for each measurements scale.⁷

Balance

In order to test the patient's overall balance, the patient was observed in several developmental positions that included tailored-sitting, quadruped, tall kneeling, half kneeling, and standing. The amount of time the patient was able to independently remain in a placed position was observed and documented at both the beginning and end of the patient's episode of care for this report. Balance was considered a significant measure since the patient's caregivers' goals included increasing the patient's independence in daily activities, and balance is required to achieve more independence with daily activities. For example, the patient requires sitting balance in order to eat or play.

Table 1: Baseline Findings & Post Treatment Results

Date	PEDI score: Functional Skills		PEDI score: Caregiver Assistance		Balance Observation: Time pt. is able to hold given position unassisted	
Week 1	Self-Care	21.4	Self-Care	0.0	Tailored-sitting	1 second
	Mobility	20.9	Mobility	0.0	Quadruped	0 seconds
	Social Function	39.6	Social Function	20.4	Tall kneeling	0 seconds
					Half-kneeling	0 seconds
					Standing	0 seconds
Week 7	Self-Care	21.4	Self-Care	0.0	Tailored-sitting	5 seconds
	Mobility	18.2	Mobility	0.0	Quadruped	0 seconds
	Social Function	37.9	Social Function	11.3	Tall kneeling	0 seconds
					Half-kneeling	0 seconds
					Standing	0 seconds

Diagnosis

The patient's diagnosis is that of Neuromuscular Practice Pattern E: Impaired Motor Function and Sensory Integrity Associated With Progressive Disorders of the Central Nervous System.⁸

Prognosis

The patient was a young male with no other significant history other than that of his diagnosis of AGS. Although the patient had a good support system and had no comorbidities, the patient was determined to have a poor prognosis for physical therapy. The progressiveness of the disease, the significant severity of the patient's symptoms, and the child's early onset of AGS determined the patient would not have a strong likelihood of rehabilitation to his level of function prior to onset of AGS.

AGS is a very progressive disorder with a poor prognosis. The prognosis of a child affected by AGS depends on the severity of symptoms and children with an earlier onset of the disorder have a higher risk of death than those with a later onset.⁴ Due to patient's severity of symptoms and early onset of the disorder, he has a much poorer prognosis than a child with a later onset. Children diagnosed with AGS often do not develop motor skills and lose any motor skills they may have acquired before onset.¹ Due to the poor prognosis, it was determined the focus of the interventions and the patient's plan of care would be to slow the patient's inevitable decline of function.

Plan of Care/ Goals

Based on the patient's caregivers' goals and the patient's significant impairments, the focus of the PT plan of care included continuing the patient's physical therapy treatment to slow the progressive decline in functional abilities. The patient received a selection of interventions proven to be effective for the child's symptoms. These interventions included neuromuscular re-education, strengthening exercises, weight-bearing activities, and passive range of movement exercises. Due to the progression of the disease, it was theorized that any positive change in the patient's functional abilities would be very minimal, if any, so measurements were taken only at the start and end of the 7-week episode of care. The patient's

caregivers were present at all sessions in order to provide carryover of treatment at home.

Over the course of 7 weeks, the patient's caregivers were asked to bring the patient for physical therapy twice a week for 60-minute sessions, with one session in a therapy pool and the second session on land at an outpatient clinic.

The short term and long term goals set for the patient are demonstrated in Table 2. Due to the patient's poor prognosis for rehabilitation as well as the progressiveness of AGS, short-term goals were set to be achieved within 6 months and long-term goals were set to be achieved within 12 months.

Table 2: Short-Term and Long-Term Goals for Physical Therapy

	Short Term Goals (6 months)	Long Term Goals (12 months)
Goal 1	Patient will increase time to independently hold a tailored-sitting position from 1 second at start of program to 5 minutes within 6 months in order to scan and engage with his environment.	Patient will increase time to independently hold a tailored-sitting position from 1 second at start of program to 10 minutes within 1 year in order to scan and engage with his environment.
Goal 2	Patient will increase his independence in ability to roll to either side from max assist x1 at start of program to mod assist within 6 months in order to more independently explore his surroundings.	Patient will increase his independence in ability to roll to either side from max assist x1 at start of program to min assist within 1 year in order to more independently explore his surroundings.
Goal 3	Patient will increase time spent with both upper extremities in midline manipulating or engaging with an object from 0 seconds at start of program to 5 minutes within 6 months in order to assist more with feedings.	Patient will increase time spent with both upper extremities in midline manipulating or engaging with an object from 0 seconds at start of program to 10 minutes within 1 year in order to assist more with feedings.
Goal 4	Patient's caregivers will be 100% compliant with Home Exercise Program in order to carryover gains from physical therapy treatment sessions and to prevent contractures.	Patient's caregivers will be 100% compliant with Home Exercise Program in order to carryover gains from physical therapy treatment sessions and to prevent contractures.

CLINICAL IMPRESSION #2

Due to the low scores on the PEDI, as well as demonstration of poor balance in all positions, it was determined that the patient should continue with interventions in order to work on achieving the goals set for the patient.

The interventions determined for the patient included neuromuscular re-education activities, strengthening exercises, weight-bearing activities, and passive range of movement exercises. All of the said interventions were done as appropriate on land and in water. Due to the lack of research on the disorder, the interventions were chosen based on clinical judgment and the

caregivers' goal of increasing the child's independence in daily activities, such as feeding and dressing, along with achieving a means of functional mobility.

INTERVENTION PROCEDURES

For the episode of care for this case report, the patient was scheduled to receive one-on-one physical therapy twice a week for 60-minute sessions, with one session in a therapy pool and the second session on land at an outpatient clinic, for 7 weeks. The patient attended 12 of the scheduled 14 visits. He missed 2 sessions during the sixth week due to going on vacation with his family for the week. The patient was not discharged at the end of the episode of care, as his goals for physical therapy had not yet been met.

Interventions were selected based on the patient's level of function and evidence in the literature. The types of interventions, as well as the rationale for the use of such interventions, are described below. Specific exercises and progressions for the 7-week treatment are listed in Appendix A. The patient's caregivers were provided a home exercise program in addition to the treatment sessions in order to maintain and build on gains.

The patient's caregivers stated that they had been consistent with his home exercise program for the course of care. At the end of the episode of care, the patient's caregivers were provided an ongoing home exercise

program and the patient was scheduled to continue with physical therapy treatments since his goals had not been met.

The patient was provided a combination of aquatic and land-based physical therapy interventions. According to a study by Getz *et al.* in 2006, aquatic interventions for children with neuromotor impairments provide several opportunities for rehabilitation that would not be possible on land. For example, the buoyancy of the water enables a child to initiate independent movements that are less likely to be achieved on land. This is significant because the child had difficulty initiating movement and the water allowed a better medium for him to work on this skill than on land. The warm temperature of a therapy pool also reduces muscle tone, which is important for this patient as he demonstrated increased spasticity secondary to influence of ATNR and STNR. It is also important to note that therapy done in a pool would also provide exercise for the patient's entire body without putting excessive stress or tension on any one area, allowing overall strengthening and conditioning for his entire body.⁹

Neuromuscular Re-education

The patient demonstrated significant delays in achieving developmental milestones as well as loss of psychomotor skills he had acquired before his early onset of AGS. Prior to onset, around 15 months of age, the patient had been independently rolling, sitting in a tailored-sitting

position during play, and was able to hold a standing position with upper extremities support at a chair or small table. Due to the progression of AGS, the patient could no longer accomplish these tasks, among many others. The loss of these skills necessitated interventions on neuromuscular re-education.

Range of Motion

The patient demonstrated some decreased gross PROM secondary to severe spasticity from the dominance of his ATNR and STNR reflexes. This impairment necessitated the use of range of motion activities in physical therapy intervention. The patient was provided passive stretching to all extremities that demonstrated spasticity during each treatment session, as this has been proven to decrease spasticity in children with spastic cerebral palsy, which presents similarly to that of a child with AGS.¹⁰

Strengthening

The patient demonstrated decreased overall strength and severe hypotonicity for all areas of the body. Therefore, the patient was a good candidate for strengthening exercises. The patient performed several task-oriented strengthening exercises including extending head and neck in order to scan his environment, kicking with his lower extremities in the pool, and picking up/ dropping toys, among others. According to a study by Salem and

Godwin in 2009, it has been demonstrated that a task-oriented strength-training program is effective for improving function in children.¹¹

Weight-bearing

Weight-bearing activities, including activities performed in supported standing positions in both land and aquatic environments, have several proven benefits to children with spastic cerebral palsy, which is similar to that of AGS. According to a study by Pin in 2007, static weight-bearing exercises with children have been shown to increase bone mineral density, thus reducing the risk of fractures. It has also been shown that static weight-bearing exercises also may temporarily reduce spasticity and may improve self esteem and communication in children as well.¹² Due to the patient's inability to stand independently to increase bone mineral density, as well as the patient's increased spasticity and difficulty in communication, it was determined that weight-bearing activities would be an appropriate part of this program.

OUTCOMES

After the 7-week course of treatment, the tests and measures were re-evaluated for the patient. The results of this re-evaluation are summarized in Table 1: Baseline Findings and Post Treatment Results. The patient demonstrated an increase in his ability to sit unsupported in a tailor-sitting

position from one second at the start of treatment to five seconds at the end of treatment. The patient demonstrated a slight decrease in a few scores on the PEDI, including the Functional Skills domains of Mobility from 20.9 to 18.2 and Social Function from 39.6 to 37.9 as well as the Caregiver Assistance domain of Social Function from 20.4 to 11.3. The other scores for the PEDI remained the same.

DISCUSSION

The purpose of this study was to examine the effectiveness of a land and aquatic-based physical therapy treatment program for improving functional limitations and balance for a 6 year-old male referred to physical therapy for developmental delay secondary to an onset of AGS. The treatments focused on neuromuscular re-education, strengthening exercises, weight-bearing activities, and PROM exercises and were chosen to decrease the symptoms of AGS and slow any decline in function as much as possible with the hope of improving the patient's quality of life.

There is little research surrounding AGS in all aspects of patient care, including physical therapy.¹ This case report contributes to the understanding of how a child with AGS may present and the impact a physical therapy program may have for a child with AGS. As more children become diagnosed with AGS each year, it is important to increase global knowledge on this progressive disorder in order to enhance awareness and

provide better care for those affected. Children can go several years without a proper diagnosis of AGS simply due to the lack of awareness of the disorder and how it presents.

Factors other than physical therapy interventions may have influenced this patient's outcome. The patient was often seen for physical therapy sessions immediately following either a speech or occupational therapy treatment session, and this may have decreased the patient's energy, focus, or attention level for treatment. The patient also would fluctuate in his performance depending on the day. For example, if the patient's caregiver was not present in the room during the entire course of treatment the patient would lose his focus and was more unwilling to participate in interventions. The patient was also not present for week six of the course of treatment, due to being on a family vacation. This may have also impacted any gains that the patient was progressing towards in treatment. It was also hard to know if the caregivers were accurate in their reports of remaining consistent with the home exercise program, and this may have also contributed to variable findings. However, the decrease in scores on the PEDI was not entirely unexpected, as AGS is a very progressive disorder and the patient would fluctuate in his performance depending on the day. Following the end of this 7-week treatment, the patient's caregivers were recommended to continue with physical therapy treatment as the patient still had not accomplished his goals for physical therapy.

In conclusion, the results from this case report demonstrated that a land and aquatic-based physical therapy treatment program may be appropriate in improving functional balance in a child with AGS, however it may not increase the patient's ability to perform overall functional activities.

Limitations of this case report may be seen in some of the data collection. There was limited data collected on tests and measures for the patient, and it may have been helpful to include other measures, such as the Modified Ashworth Scale which measures a patient's spasticity, or perhaps a survey that measured the caregivers' satisfaction with the course of treatment or a measure that looked more at the patient and/or caregivers' quality of life.¹³ It would have also been beneficial to have had a longer time frame to collect more data in order to see a clearer change in function since AGS is so progressive and change would be minimal, if any, in such a short time frame.

Suggestions for future research include a longitudinal study of physical therapy intervention beginning immediately following diagnosis of AGS since AGS is a very progressive disorder and children often live several years with the disorder. Studies relating AGS to other similar diagnoses, such as cerebral palsy, spinal muscular atrophy, or Rett syndrome would also be beneficial.

References

1. Juern A, Robbins A, Galbraith S, Drolet B. Aicardi-Goutières Syndrome: Cutaneous, Laboratory, and Radiologic Findings: A Case Report. *Pediatric Dermatology*. January 2010;27(1):82-85. Available from: Academic Search Complete, Ipswich, MA. Accessed October 28, 2014.
2. Rossler L, Ludwig-Seibold C, Thiels C, Schaper J. Aicardi-Goutières syndrome with emphasis on sonographic features in infancy. *Pediatric Radiology*. August 2012;42(8):932-940. Available from: CINAHL Complete, Ipswich, MA. Accessed October 28, 2014.
3. Chahwan C, Chahwan R. Aicardi-Goutieres syndrome: from patients to genes and beyond. *Clinical Genetics*. May 2012;81 (5):413-420. Available from: Academic Search Complete, Ipswich, MA. Accessed October 28, 2014.
4. National Institute of Neurological Disorders and Stroke (NINDS). National Institute of Health. NINDS Aicardi-Goutieres Syndrome Disorder Information Page. *NIH: National Institute of Neurological Disorders and Stroke*. 18 November, 2010. Available at http://www.ninds.nih.gov/disorders/aicardi_goutieres/aicardi_goutieres.htm. Accessed October 28, 2014.
5. Berg M, Jahnsen R, Frøslie K, Hussain A. Reliability of the Pediatric Evaluation of Disability Inventory (PEDI). *Physical & Occupational Therapy In Pediatrics*. September 2004;24(3):61-77. Available from: Academic Search Complete, Ipswich, MA. Accessed October 28, 2014.
6. Ragnarsdottir M. The concept of balance. *Physiotherapy*. June 1996;82(6):368-375. Available from: CINAHL Complete, Ipswich, MA. Accessed October 28, 2014.
7. Feldman AB, Haley M, Coryell J. Concurrent and construct validity of the Pediatric Evaluation of Disability Inventory. *Phys Ther*. 1990;70: 602-610.
8. APTA Guide to Physical Therapist Practice. Second Edition. American Physical Therapy Association; *Phys Ther*. Jan 2001; 81(1): 305
9. Getz M, Hutzler Y, Vermeer A. Effects of aquatic interventions in children with neuromotor impairments: a systematic review of the literature. *Clinical Rehabilitation*. November 2006;20(11):927-936. Available from: Academic Search Complete, Ipswich, MA. Accessed October 28, 2014.
10. Tamis P, Paula D, Michael C. The effectiveness of passive stretching in children with cerebral palsy. *Developmental Medicine & Child Neurology*. October 2006;48(10):855-862. Available from: Academic Search Complete, Ipswich, MA. Accessed October 28, 2014.
11. Salem Y, Godwin E. Effects of task-oriented training on mobility function in children with cerebral palsy. *Neurorehabilitation*. June

- 2009;24(4):307-313. Available from: Academic Search Complete, Ipswich, MA. Accessed October 28, 2014.
12. Pin T. Effectiveness of static weight-bearing exercises in children with cerebral palsy [published erratum appears in PEDIATR PHYS THER 2007 summer;19(2):172]. *Pediatric Physical Therapy*. March 2007;19(1):62-73. Available from: CINAHL Complete, Ipswich, MA. Accessed October 28, 2014.
13. Ansari N, Naghdi S, Mashayekhi M, Hasson S, Fakhari Z, Jalaie S. Intra-rater reliability of the Modified Modified Ashworth Scale (MMAS) in the assessment of upper-limb muscle spasticity. *Neurorehabilitation*. July 2012;31(2):215-222. Available from: Academic Search Complete, Ipswich, MA. Accessed October 28, 2014.

Appendix A: Comprehensive Plan of Care

Week #	Neuromuscular re-education	Strengthening exercises	Weight-bearing activity	ROM exercise
1	-“stepping” movements platform in pool and on land -PNF to UE’s and LE’s in pool -placement into prone-on-elbows position to encourage neck/head extension in both pool and land (floating mat used in pool)	-kicking LE’s in pool to “swim” -picking up soft ball and raising arms to drop in ring on land	-standing on platform in pool while playing with toys with UE’s	-passive stretching to bilateral ankles in pool and on land
2	-“stepping” movements on platform in pool and on land -PNF to UE’s and LE’s in pool -placement into prone-on-elbows position to encourage neck/head extension in both pool and land (floating mat used in pool)	-kicking LE’s in pool to “swim” -using arms to help push trunk upright when in supported sitting	-standing on platform in pool while playing with toys with UE’s -placed into supported half kneeling position during play	-passive stretching to bilateral ankles in pool and on land
3	-“stepping” movements on platform in pool and on land -PNF to UE’s and LE’s in pool -kinesiotape provided to neck extensors for reminder to keep “head up” -placement into prone-on-elbows position to encourage neck/head extension in both pool and land (floating mat used in pool)	-kicking LE’s in pool to “swim” -placed in supported sitting position with arms extended and encouraged to extend head/neck to see bubbles	-standing on platform in pool while playing with toys with UE’s -placed into supported tall kneeling position during play	-passive stretching to bilateral ankles in pool and on land
4	-“stepping” movements on platform in pool and on land -reaching for objects while on side of pool while supported in water -reaching for objects while in prone-on-elbows position on land	-kicking LE’s in pool to “swim” -moving in/out of hip flex/ext while in supported kneeling/prone-on-elbows position with PT support	-standing on platform in pool while playing with toys with UE’s -standing at table while using UE’s to play with rolling a ball	-passive stretching to bilateral ankles in pool and on land
5	-“stepping” movements on platform in pool and on land -tapping shovel on bucket in pool with UE’s -assisted rolling supine<->prone	-kicking LE’s in pool to “swim” -pushing off of wall in pool with LE’s in a supported supine position -work on extension/flexion of UE/LE while prone on land and pool	-standing on platform in pool while playing with toys with UE’s -standing at table while using UE’s to play with car set	-passive stretching to bilateral ankles in pool and on land
6	N/A: vacation	N/A: vacation	N/A: vacation	N/A: vacation
7	-“stepping” movements on platform in pool and on land -lite-gait equipment used to help with “stepping” movements on land -assisted rolling supine<->prone on land	-kicking LE’s in pool to “swim” -pushing off of wall in pool with LE’s in a supported supine position -using lite-gait to work on “kicking” a soft ball with LE’s	-standing on platform in pool while playing with toys with UE’s -standing in lite-gait	-passive stretching to bilateral ankles in pool and on land