Case Study: Unilateral Schizencephaly-How Resistance Training Affects EMG Signal and Muscle Strength

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Abstract

Introduction: To date, there is limited research showing how skeletal muscle in individuals with unilateral schizencephaly responds to traditional weight training programs. In all cases of schizencephaly motor impairment is by far the most frequent deficit. Examining strength increases that could be achieved in individuals with unilateral schizencephaly has clinical, as well as rehabilitative potential to help these individuals achieve a wider range of muscular independence, thereby improving their quality of life. It is hypothesized that skeletal muscles affected by unilateral schizencephaly will see a similar percent strength gain (torque production) when compared to unaffected skeletal muscles after an 8-week strength training program. Furthermore, it is estimated that the skeletal muscles affected by unilateral schizencephaly will exhibit a greater percent increase in electromyography (EMG) signal activity after the 8-week training program as compared to the unaffected contralateral skeletal muscles. Methods: The participant underwent an 8-week periodized unilateral independent lifting protocol performed on machine and cable weights. During the 8-week program, a pre-test, mid-test, and post-test were administered using an isokinetic BioDex protocol and Electromyography system to measure functional muscle strength as well as muscle stimulation. Results: EMG signal increased for the left (affected) biceps brachii muscle as well as the vastus lateralis and medialis muscles in the left (affected) leg. No increase was shown for right biceps brachii muscle, right vastus lateralis muscle, right vastus medialis muscle, as well as right and left rectus femoris muscles. Average peak torque increased for the participants left (affected) leg and right (unaffected) arm, yet remained constant or even declined slightly for the right (unaffected) leg and left (affected) arm. Conclusion: Observation of skeletal muscle affected by unilateral schizencephaly shows a positive response to strength training. It is proposed that future studies should be conducted to investigate how strength training more specifically affects unilateral schizencephaly on 1) gross motor function/gait analysis/range of motion, 2) neurological function.

Key Words: cerebral cortex, skeletal muscle, strength training
Introduction
Schizencephaly is a rare developmental malformation of the cerebral cortex. Recent estimates show that the rate of incidence of schizencephaly is reported as being 1.54/100,000.1 Specifically, schizencephaly is characterized by the formation of clefts or slits in the cerebral cortex during prenatal development. While the precise pathogenesis of schizencephaly is a matter of debate, a commonly accepted theory argues that vascular damage in the early stages of neuroembryogenesis results in vascular deficiency to an area of the germinal matrix, inhibiting neuronal development and leading to the formation of a cleft.2 These irregular clefts in the brain extend from the surface of the pia mater down into the cerebral ventricles. The margins in the cleft are then lined with heterotrophic grey matter.3 These clefts in the cerebral cortex of affected individuals interfere with the development and further functioning of body systems. Clinical manifestations include motor deficits and mental retardation, but the severity of these deficits is extremely variable; mainly related to the size and location of the clefts.4 Schizencephaly can be manifested unilaterally, affecting only one side of the cerebral cortex, or bilaterally affecting both sides. In both types of schizencephaly motor impairment is by far the most common side effect.4

The lack of motor control leads to dramatic muscle atrophy and weakness on the affected side(s) of the body. Theories have speculated that the motor processes of damaged motor cortex are altered or assumed by neurons in healthy brain tissue due to the plasticity of the brain, thereby compensating for deficiencies in damaged or diseased brain.5 Such assumptions were verified by Belardinelli et al. (2007) who showed that the intact hemisphere assumes motor control of the paretic ipsilateral hand.

Current research has focused on developmental components of this malformation. There is limited research showing how skeletal muscle in individuals with schizencephaly respond to traditional weight training programs. It is known that in clinical trials studying Cerebral Palsy (CP), a malformation closely related to schizencephaly, muscle weakness contributes to muscular imbalance (agonist vs. antagonist) with gross motor skills. A relationship has also been shown between muscle weakness and finer motor skills such as reaching and grasping.6 This imbalance can lead to further weakness, atrophy, and joint contracture.7 These functional limitations impede normal development and age-appropriate skills.

Traditional treatment of children with CP has tended to focus on mitigating spasticity, which was thought to exacerbated with exercise, as opposed to mitigating muscle weakness.8 In the late 1990’s research began to show that muscular strength training exercises were impactful in improving lower extremity joint strength,
walking speed/cadence, and gross motor function. Damaino et al. (2002) showed that strength exercises were indeed beneficial in improving muscular strength and gait functions in children with CP. In 2012 Lee et al. used the Comprehensive Hand Repetitive Intensive Strengthening Training (CHRIST) method to report improvement in functional motor performance of the upper extremity as well as increased muscle size in a patient with bilateral schizencephaly. Given that strength training has been shown to have beneficial effects on both upper and lower extremity gross and fine motor skills in children with CP and bilateral schizencephaly, the researchers of this study sought to examine the effect that strength training would have on individuals with unilateral schizencephaly. There is clinical, as well as rehabilitative potential to help these individuals achieve a wider range of muscular independence, subsequently improving their quality of life. As seen in cases of CP, there may be a comparable response of skeletal muscle between the unaffected and affected muscles. Therefore, the researchers are optimistic that a well-structured training program targeting major primary movers, as well as minor support and stabilizing muscles will yield substantial musculoskeletal improvements, increased function, and range of motion leading to improved quality of life.

Recent analysis of patients with CP suggest that motor-unit firing rates of lower extremity muscles do not differ from healthy individuals. This appears to be, in part, due to the inability of damaged motor pathways to provide sufficient excitatory impulses to fully activate the entirety of available motor units. Rose et al. suggest that interventions aimed at building strength may be effective for reducing movement deficit and improving gait in that they increase voluntary excitatory drive and muscle activation. As it relates to unilateral schizencephaly, it is possible that damaged motor pathways are, at least in part, responsible for movement deficits or abnormalities demonstrated by the participant. The researchers of this study postulate that increased EMG signal will be seen following a strength training regime in the affected muscles as a result of increased excitatory drive to the local motor neurons.

The aim of this study is to examine the response of affected skeletal muscles to strength training in a participant with unilateral schizencephaly. A secondary aim of this study is to examine the motor-activation signal changes, as observed through electromyography (EMG) data, that occur following an 8-week exercise program.

It is hypothesized that skeletal muscles affected by unilateral schizencephaly will see a similar percent strength gain (torque production) when compared to the skeletal muscles that are not affected by the disorder following an 8-week strength training program. Furthermore, it is
estimated that the skeletal muscles affected by unilateral schizencephaly will exhibit a greater percent increase in electromyography (EMG) activity after the 8-week training program as compared to the unaffected skeletal muscles.

**Methods**

**Participant**
The participant in this study was a 22-year-old untrained male, 1.75m tall, 77.1 kg., with a relatively less severe case of unilateral schizencephaly. The participant did not exhibit any other disorders that are commonly found to coexist with this malformation. Physical manifestations of this participant include atrophied skeletal muscle, bone, and soft tissue in his left extremities. He did not present with additional disorders, which is of benefit to this study in that there are no other physical or mental limitations that need to be taken into consideration while collecting data. Unilateral schizencephaly can be observed and tested without confounding factors from these other common disorders.

**Experimental design**
To test force production of the muscle groups in question, a custom isokinetic unilateral protocol was created utilizing the BioDex (BioDex, Inc.; Shirley, NY) system. EMG (Delsys Inc.; Natick, MA) data were recorded throughout the BioDex testing. The participant was outfitted with eight wireless EMG sensors; one located on each muscle in question (left and right biceps brachii m., vastus lateralis m., rectus femoris m., vastus medialis m.). The first step of the protocol was gathering maximal voluntary isometric contraction (MVC) EMG data for every muscle being observed. The BioDex dynamometer was positioned for optimal elbow and knee joint angles and MVC EMG data were collected. Two MVC’s were performed for each EMG muscle sensor, and a mean value was calculated for the MVC.

MVC’s were administered immediately prior to the participant performing each individual joint protocol on the BioDex dynamometer. The custom isokinetic unilateral BioDex protocol was administered to the participant and consisted on four sets of five reps for each muscle group being tested. Each set progressively decreased the controlled angular velocity of the BioDex dynamometer from 240 °/s, 180 °/s, 120 °/s, and 60 °/s during the working phase. The angular velocity of the BioDex dynamometer during relax phase was consistent at 300 °/s for all joints, simulating essentially unrestricted range of motion. EMG data were recorded for all isokinetic exercises. The order in which muscles were tested were right bicep brachii muscle, left bicep brachii muscle, right quadriceps, and left quadriceps.

**Participant exercise**
The participant completed an eight-week weight-training program designed by the researchers to increase strength in the
affected muscles. The participant exercised three times each week for eight weeks in the Student Life and Wellness building at Utah Valley University. The exercises that the participant performed included (in exercise order): chest press, biceps curl, triceps extension, seated rows, leg extension, shoulder press, hamstring curl, lat pull, leg press, and core plank. The participant was allowed to rest for approximately 30 seconds in between each set of exercise and approximately 2 minutes during the transition to each new exercise. Each training session was approximately one hour in duration and was supervised by the researchers. All exercises were performed on weight machines manufactured by LifeFitness (LifeFitness, Inc., Rosemont, IL). To promote healthy strength increase, the exercise program was designed to utilize movements from all major muscle groups.

The researchers traditionally periodized the 8-week program. Traditional periodization promotes wave-like progression (periods of overload interspersed with periods of recovery), typically moving from general training (high volume/low intensity) towards specific training (low volume/high intensity). The periodized program for this study was constructed of two meso-cycles with a mid-test on the EMG/BioDex protocols administered at the end of the first meso-cycle. Each meso-cycle was further divided into 4 micro-cycles, corresponding with each successive week of exercise. A baseline test was administered to determine starting weights for the right (unaffected) side and left (affected) side his body. The researchers then calculated a 10% increase of weight for each exercise each week; with a return to the designated weight for micro-cycle one during micro-cycle four of each meso-cycle. This drop in intensity was done the week prior to the mid-and-post EMG/BioDex tests in order to see peak performance (see Table 1).

Results
All procedures were successfully carried out with the participant. With the view toward successfully identifying the response of skeletal muscle affected by unilateral schizencephaly to strength training, data were obtained from our participant for each muscle group in question according to our above mentioned methods (Table 2).

In consideration of the three left knee extensors examined, EMG increased signal percent output was observed in the vastus medialis muscle (16.37% ± 4.1%), and vastus lateralis muscle (8.3% ± 2.17%). However, the left rectus femoris muscle saw a decline in overall EMG activation of (-3.35% ± 1.1%) [see Figure 1]. In consideration of the left biceps brachii muscle, the largest percent increase of EMG activation was recorded (24.95% ± 1.8%) [see Figure 1].

An examination of the data obtained for the participant’s right (unaffected) extremities showed a general decline of overall EMG activity from the pre-test to the post-test. In
consideration of the right leg knee extensors examined, a decrease of EMG activity was seen for the vastus medialis muscle (-1.92% ± 1.2%) and vastus lateralis muscle (-16.17% ± 1.9%). However, the right rectus femoris muscle saw a decline in overall EMG activation of (-22.56% ± 1.2%) [see Figure 2]. In consideration of the right biceps brachii muscle, a slight decrease in EMG activation was recorded (-0.1% ± 6.5%) [see Figure 2].

Table 1. Traditionally periodized Exercise Program for a Participant with unilateral schizencephaly.

<table>
<thead>
<tr>
<th>Micro-Cycle</th>
<th>Meso-Cycle #1</th>
<th>Meso-Cycle #2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre-Test</td>
<td>1  2  3  4</td>
<td>5  6  7  8</td>
</tr>
<tr>
<td>Post-Test</td>
<td></td>
<td>Post-Test</td>
</tr>
</tbody>
</table>

Table 2: Mean Peak Torque and standard deviation of each joint tested during an 8-week strength training program in an individual with unilateral schizencephaly.

<table>
<thead>
<tr>
<th>Average Peak Torque (ft*lb)</th>
<th>Pre-Test</th>
<th>Mid-Test</th>
<th>Post-Test</th>
<th>% Change</th>
<th>Pre-Test</th>
<th>Mid-Test</th>
<th>Post-Test</th>
<th>% Change</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Left Knee</td>
<td>Left Elbow</td>
<td></td>
<td></td>
<td>Right Knee</td>
<td>Right Elbow</td>
</tr>
<tr>
<td>240 °/s</td>
<td>44.6</td>
<td>54.6</td>
<td>59.3</td>
<td>24.80%</td>
<td>13.4</td>
<td>14.2</td>
<td>14.5</td>
<td>7.60%</td>
</tr>
<tr>
<td>180 °/s</td>
<td>47.4</td>
<td>58.2</td>
<td>64.9</td>
<td>27.00%</td>
<td>14.3</td>
<td>13.9</td>
<td>12.8</td>
<td>-11.70%</td>
</tr>
<tr>
<td>120 °/s</td>
<td>61.8</td>
<td>75.6</td>
<td>77.7</td>
<td>20.50%</td>
<td>13.9</td>
<td>14.1</td>
<td>12.2</td>
<td>-13.90%</td>
</tr>
<tr>
<td>60 °/s</td>
<td>84.4</td>
<td>92.9</td>
<td>101</td>
<td>20.50%</td>
<td>13.9</td>
<td>14.1</td>
<td>12.2</td>
<td>-13.90%</td>
</tr>
</tbody>
</table>

| 240 °/s                    | 106.9    | 98.2     | 93.9      | -13.80%   | 21.7     | 32.2     | 27        | 19.60%    |
| 180 °/s                    | 123.1    | 107.3    | 103.1     | -19.40%   | 24.7     | 34.4     | 31        | 20.30%    |
| 120 °/s                    | 140.8    | 130.1    | 132.2     | -6.50%    | 27.8     | 35.5     | 33.5      | 16.80%    |
| 60 °/s                     | 155.8    | 142      | 151.3     | -3.00%    | 30       | 40.5     | 34.4      | 12.80%    |
Electromyography signaling of the observed left (L) upper and lower extremity muscles in a participant with unilateral schizencephaly during an 8 week strength training program.

**Figure 1.** Electromyography signals for the observed (L) upper and lower extremity muscles.

Electromyography signaling of the observed right (R) upper and lower extremity muscles in a participant with unilateral schizencephaly during an 8 week strength training program.

**Figure 2.** Electromyography signals for the observed (R) upper and lower extremity muscles.
Discussion

It was hypothesized that after an eight-week strength training program skeletal muscles affected by unilateral schizencephaly would have a similar percent strength gain (torque production) when compared to the skeletal muscles that are not affected by the disorder. This case-study found that following an eight-week strength training program skeletal muscle affected by unilateral schizencephaly showed an increase of torque production about the knee and elbow joint. Contrary to the hypothesis, the increase of torque production of the affected side vs. the unaffected side were unequal, and will be discussed further. Furthermore, it was estimated that affected skeletal muscles would exhibit a greater percent increase in EMG activity after the eight-week training program as compared to the unaffected skeletal muscles. This study found that following an eight-week strength training program, affected muscles demonstrated an increase of EMG signal strength.

This study, of unilateral schizencephaly, found similar results to Lee et al.; that upper extremity muscular strength and motor control could be improved in patients with cerebral cortex malformations through strength training.\textsuperscript{11}

Upper Extremity vs. Lower Extremity

The participant’s left leg saw a decrease in EMG signal of the rectus femoris muscle and an increase in EMG signal of the vastus lateralis and vastus medialis muscles [see Figure 1]. This could be attributed to increased agonist muscle function derived from strengthening the left quadriceps muscle group. Increased muscle coordination could be the result of the strength-training program completed by the participant. When healthy individuals move the knee in to extension, increased coordination of the quadriceps muscle group allows for a more balanced recruitment of needed muscles to perform the given extension.

It is undetermined why a decrease in EMG signals in the right leg was shown following training [see Figure 2]. It has been shown in the literature that during the development of gait in persons with CP, the musculoskeletal resources that are available to the person are utilized to develop an optimal gait pattern in the absence of normal muscle forcing.\textsuperscript{15} It is possible that through adolescent development the participant fostered a reliance on his right side (unaffected) musculature to complete many activities of daily living such as ambulation. Now that the participant has reached adulthood, many of his modified activities of daily living have become habit. While the training did not diminish this reliance completely, it may have adjusted it slightly. It could be that with increased strength and ability in his left (affected) side, the workload demand on the right side decreased resulting in negative strength adaptations.
EMG and BioDex data of the lower extremities support each other, finding that the affected (left) side saw greater increases in torque production and muscle stimulation. However, EMG and BioDex data of the upper extremities showed inconsistent results between left and right biceps brachii muscles. The right (unaffected) side showed an increase in muscle strength, but no change in EMG signals. The researchers speculate that this may be due to normal neurological adaptations seen in weight training. As the muscle increases in strength, neurological adaptations that lead to the decreased signals from the CNS to the working muscles may include a decrease in inhibitory postsynaptic action potential from muscle proprioceptors and increases in coordination of motor unit recruitment. Baechle states that an important consideration, when collecting EMG data, is the training status of the individual. This is because neural adaptations predominantly occur early in training and lead to improved motor learning and coordination. Given that overall, the participant was untrained, the fact that his right arm is unaffected by the malformation indicates that his right arm may have been in a naturally more trained state than to his left arm at the onset of this study. The disparity between the participant’s left and right upper extremity EMG increases is supported by previous research which states 30 percent of studies that show an increase in muscle strength and power do not show changes in EMG. It is believed that the lower body’s unaffected side (right) did not see these results because it is highly cooperative with the (left) affected leg in ambulation and will respond differently as the affected leg increases in strength.

Upper Extremity Comparisons
By way of comparison between upper and lower extremity muscle systems, a larger increase of EMG signal was observed in the participant's affected (left) upper extremity. It was seen that the left biceps brachii muscle had larger EMG signal gain over the course of the training program as compared to both left and right quadriceps muscle groups [see Figures 1,2]. The left arm also showed a greater increase in EMG signal than the right arm. This observed difference between EMG signal increases of the lower and upper extremity could be normal based on the difference in activity level between the two. The quadriceps muscles are regularly stimulated during activities of daily living; such as ambulation, postural support, and weight bearing exercise. The biceps brachii muscles, in contrast, are not used regularly in weight bearing activities, which result in less stimulation when compared to lower body muscle groups. This allowed the upper body a greater potential for improvement. In light of the limited duration of this present study, an 8-week strength program, the researchers postulate that the initial adaptations observed in the upper extremities were steeper because of this greater potential for improvement. It would
be expected that if strength training continued beyond eight weeks the rate of improvement of the biceps brachii muscles would decrease based on the law of diminishing returns.

During the exercise program, the participant experienced pain in his left (affected) shoulder joint when performing seated biceps curls on a machine that prohibited free range of motion. It was observed that the participant developed a more medially oriented carry-angle between his left shoulder and elbow in order to maximize the kinetic leverage of his left arm. Though this medially oriented carry-angle was not physically measured, it is worth noting that it was visually noticed by the researchers when the subject drew his brachium toward his torso during repetitions of the seated biceps curl. This pain is believed to be a direct result of the abnormal carry-angle that was developed. To mitigate the shoulder joint pain, the researchers instructed the participant to abduct his arm so it did not rest on the supportive elbow pad of the bicep curl machine resulting in a more isolated, pain free movement during the exercise. The researchers postulate that this abnormal carry-angle affected data collection during the pre, mid, and posttests with the BioDex dynamometer because the dynamometer restricts the elbow to a pre-specified sagittal plane motion. The BioDex dynamometer does not permit any type of freedom for varus/valgus deviations during elbow joint flexion/extension range of motion. The researchers suggest that though increases in the EMG signal of the left biceps brachii muscle were observed, no increases torque production were observed because of the limitations that the BioDex dynamometer placed on his adapted range of motion.

**Cross-Education Effect**

During the strength-training program, the researchers were intrigued by the observation of the participant contracting the contralateral affected (left) muscles involuntarily and simultaneously with the contraction of the non-affected (right) muscles. It has been reported that chronic unilateral motor activity can affect homologous muscles in the contralateral limb. The researchers speculate that the participant’s observed involuntary contraction may be attributed to the left cerebral hemisphere developing the ability to compensate for the diminished motor control on the affected left side. From a developmental perspective, it is likely that motor neurons with origins in the left cerebral cortex adapted and “crossed-over” to assist the affected right motor cortex achieve a higher level of operation. It is highly likely that the left side of his brain has adapted such to have some control over the left side of his body as well as controlling the right side as it normally would.

Additionally, an increased EMG signal could be explained considering the mirroring effect inherent in individuals with cerebral
cortex malformations, where the unaffected cortex assumes the duties of the affected in an attempt to compensate for resulting deficiency. Given that two hemispheres of the brain are now involved in producing an action potential in the left bicep, and that the aforementioned muscle is unaccustomed to frequent functional stimulation, it is our belief that the notable increase observed in left bicep EMG activation is the result of the brain learning to activate all appropriate motor units in the correct sequence and not necessarily strength increases.

It stands to reason that the participant began developing this compensatory mechanism early in childhood, quite possibly as early as the prenatal developmental stage, and into the beginning stages of post-natal motor development. Such investigation is beyond the scope of this study. However, the researchers were provided with information about the participant’s development as a child; including that he did not begin walking until he was four years old and, even then, required a brace on the left leg to aid in straightening the foot.

**Discussion of Exercise Program**

During each exercise bout, the participant’s left (affected) wrist was in a constant state of flexion -- what appears to be Volkmann’s contracture. This affected his grip strength and stability during the cable rows, seated bicep curls, and lat pulldown exercises. It was suggested to the participant that he extend the wrist of the opposing unaffected limb. This, surprisingly, helped straighten out the affected wrist, decreasing the effects from the Volkmann’s contracture. This mirroring supports the idea that the unaffected hemisphere is attempting to compensate for the lack of stimulus from the affected motor cortex. More research would need to be done in order to determine if the mirroring effect progressively diminishes with prolonged unilateral independent resistant training.

After measuring the participant's 10-rep max on the first day of exercise, he became light headed and dizzy. To ensure the complete safety of the participant, the workout was immediately terminated. The participant was then accompanied for 30 minutes and provided with food and water. The participant was then advised to eat a balanced meal at approximately 60 minutes prior to exercise sessions. For subsequent exercise bouts, his adherence to this advice greatly enhanced his energy level and eliminated dizziness and lightheadedness.

**Limitations**

It should be noted that potential limitations to this study could have decreased the accuracy of the findings. Such limitations include scheduling conflicts with our participant, making the strength-training program less consistent than expected, as well as occasionally having to decrease the duration of each exercise bout. To mitigate the impact of scheduling conflicts on participant exercise, significant effort was
made by the researchers to provide adequate alternative times for the participant to schedule exercise. This enabled the participant to minimize missed exercise time.

While there were increases and decreases between the pre- and mid-test, and the mid- and post-test EMG signals, the researchers believe that the initial responses, that is, those taking place between the pre- and mid-test, were resultant of the participant’s involvement in a new exercise regimen. They also postulate that those responses taking place between the mid- and post-test were largely the result of decreased participant involvement in regularly scheduled exercise.

**Conclusion**

Observation of skeletal muscle affected by unilateral schizencephaly shows a positive response to strength training. Substantial strength increases were seen in consideration of average torque production as well as EMG activity for the affected (left extremity) skeletal muscle. Implementation of these findings carry clinical as well as real-world significance. For those with affected by unilateral schizencephaly, training programs can be designed to improve the dexterity and operation affected limbs. This in turn can help patients achieve greater independence and quality of life by increasing their physical capabilities. The researchers propose that future studies be conducted to investigate how strength training more specifically affects unilateral schizencephaly on 1) gross motor function, gait analysis, and ROM, 2) neurological function.

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**References**


