# PHYSIOTHERAPY MANAGEMENT OF THE SICKLE CELL HEMIPLEGIC PATIENT — A Case Report

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*ABSTRACT*
*Sickle cell anaemia patients commonly have several complications that may negatively affect their health and quality of life. Treatment of this condition usually involves different experts in the medical field. This case report evaluates the effect of electrical stimulation in conjunction with other physiotherapy modalities in managing spasticity secondary to cerebrovascular accident in a sickle cell anaemia patient.*

*The findings in this case report showed that electrical stimulation did not aggravate spasticity when used in conjunction with other physiotherapy modalities.*

**Key words:** cerebrovascular accident, spasticity, atrophy, electrical stimulation

## INTRODUCTION

Sickle cell is a term used to denote a group of inherited disorders occurring mostly in African people or people of African origin. The disorder is characterized by a predominance of haemoglobin-S (HbS) in the red blood cell.[1] Sickle cell disease (SCD) has been observed to be fatal in childhood.[2] The substitution of valine for glutamic acid in the sixth position of the amino acid sequence of the beta chain results in the locking of the adjacent ends of the alpha chains. Following this, the haemoglobin molecules become stacked in rows. This results in the distortion of red blood cells to a sickle shape in deoxygenated blood. The sickling can be demonstrated by adding a reducing agent to the blood.[2]

Clinically, in sickle cell disease, there is chronic haemolytic anaemia, painful sickling crises, leg ulceration, recurrent respiratory infection, myocardial insufficiency and various vaso-occlusive episodes. Hemiplegia in sickle cell disease is usually a result of the occlusion of arterial blood supply to the brain. Cerebrovascular accident (CVA) has been observed to be one of the complications usually associated with sickle cell disease. The devastating effect of the residual paralysis on a child cannot be quantified, hence, adequate rehabilitative programmes should be designed to ensure that such hemiplegic children live fulfilled lives. Associated complications of neurological damage include: shoulder subluxation, loss of functional ability in the hands, foot drop and psychological trauma.

The place of electrical stimulation in the management of hemiplegic patients has remained controversial. Various investigators have, however, established the importance of electrical stimulation in the prevention/management of glenohumeral subluxation and atrophy in hemiplegic patients.[3,4,5] Also, the belief that electrical muscle stimulation exacerbates spasticity is gradually being eroded. No known previous studies have reported an increment in spasticity following the use of electrical stimulation.

There are three different schools of thought in the management of spasticity using electrical stimulation. They are those who believe in (1) stimulation of the antagonist muscle; (2) direct stimulation of the spastic muscles; and (3) alternate stimulation of the agonist and antagonist muscles. The last approach has been tried using low frequency (3-35HZ) 0.2ms pulse duration for some minutes daily over several weeks.[6] The principle behind this is that afferent stimulation reinforces the presynaptic inhibition of motor neurons, thus reducing spasticity. Largesse and Loy[5] studied the effect of functional electrical stimulation on the co-contraction level of a spastic hemiparetic patient and observed that there was a reduction in the antagonist co-contraction level

associated with spasticity. Another study, however, found only short-lived improvement in spasticity.

Similarly, faradic stimulation has been observed to be a good replacement for splinting. For example, electrical stimulation of the dorsiflexors in the management of patients with foot drop and in the prevention of glenohumeral subluxation in hemiplegic patients has been observed to be beneficial.

To the best of my knowledge, there is no known previous study of the use of electrical stimulation in the management of a sickle cell hemiplegic patient. This case report was therefore carried out to fill this vacuum.

## METHOD

### Subject

RB is a 13 year-old girl who reported at our hospital on the 7th of April 1999, with a two-day history of bone pain in all her limbs and loss of functional ability in the left limbs. She also reported twitching of her left facial muscles followed by deviation to the right. Her mother reported no history of trauma.

### Past Medical History

RB was diagnosed as an SCD patient at the age of 3 years. She also had a history of recurrent malaria and hospitalization on 3 different occasions. She had been given two doses of dexamethasone at a private hospital before reporting at our hospital. RB had previously had an exchange blood transfusion (EBT); the details of this could not be obtained.

### Family and Social History

RB is the sixth of seven children and the only living sickler. The other two older sicklers are dead. She is a secondary school student.

### Physical Examination

On examination, she looked ill, was afebrile, jaundiced$^{++}$, not distressed, had no pedal oedema, no significant lymphadenopathy and had a good hydration status. She was conscious and alert, well oriented in TPP (time, place and person).

**Chest.** .Respiratory rate (RR) 28 breaths/min. Clinically clear with normal and full thoracic excursion.

**Cardiovascular System (CVS).** BP was 120/75mmHg, pulse was 98b/mins regular and of good volume; auscultation, however, revealed haemic murmur.

**Abdomen.** Full, soft with no tenderness.

**Limbs.** Had passive full range of motion but was globally hypotonic with a left gross muscle power of zero in both the left upper and lower limb muscles. There was no obvious muscle atrophy in all the muscle groups and there was good bone alignment in the left shoulder joint.

**Reflexes.** She exhibited increased biceps tendon and deep knee tendon reflexes. Similarly, ankle clonus and Babinski reflex were present. These were major signs of upper motor neuron lesion.

**Neurological assessment:** Her skin sensation was observed to be globally intact and Kernig's sign was negative (absent). Her grade (see appendix) on Bronstom assessment was zero.

Following the examination, right cerebrovascular accident (CVA) secondary to a vaso-occlusive episode was diagnosed. Also, the result of the blood culture showed no growth after 7 days incubation and urinalysis was observed to be within the normal range. From the laboratory result, conjugated hyperbilirubinemia was diagnosed because her conjugate bilirubin level was found to be high - 18 micromol/litre (normal range is 0-2 micromol/litre).

The management plan designed for RB on admission was: intravenous fluid 4.3% in $^1/_{5}$th saline, subcutaneous chloroquine injection 5mg/kg 12 hourly x 3 doses. She was placed on an anticoagulant therapy in the form of Aspirin 75mg daily.

## PHYSIOTHERAPY MANAGEMENT

Physiotherapy management commenced after RB was admitted to the Emergency Pediatric Unit (EPU). For the first 3 weeks, management was limited to a bed programme in the ward because the patient was generally weak and could not withstand frequent transfer. The programme consisted of passive movement, proprioceptive neuromuscular facilitation, positioning and regular turning three times per day. Four weeks post-admission, she developed moderately severe elbow flexor spasticity and pain in the left shoulder, knee and ankle joints.

The management therapy also included infra-red radiation using a tunnel bath (20 minutes each for the left upper and lower limbs), electrical stimulation (ES) through interrupted direct current (EMS-Myodine 5 electrical simulator) with 66 volt, 300ms, rectangular pulse duration for 10 minutes each to the left deltoid (anterior, middle and posterior fibres) elbow flexors, triceps, left facial muscles, quadriceps femoris and ankle dorsiflexors. In addition, general

mat exercises and standing re-education with the aid of a back slab commenced.

The upper limb (i.e., left elbow flexors) received ES directly while the left quadriceps muscles which serve as the antagonist for the left knee flexors were stimulated.

## RESULT

Seven weeks after the commencement of physiotherapy management in the department, the patient was re-assessed and was found to have improved in gross muscle power (left upper limb -2 and left lower limb - 3). Table 1 shows the result of the selective muscle power testing. She was also able to ambulate within the parallel bar satisfactorily. The grade on Bronstom assessment was 2 (see appendix).

**Table 1.** Muscle assessment * chart for the left upper and lower limbs

| Muscles | Muscle power on admission | Muscle power 7 weeks post admission | Muscle power 11 weeks post admission |
|---|---|---|---|
| Shoulder flexors | 0 | 2 | 3 |
| Shoulder abductors | 0 | 2+ | 3 |
| Shoulder extensors | 0 | 2 | 3 |
| Elbows flexors | 0 | 2 | 3 |
| Elbows extensors | 0 | 2+ | 3+ |
| Wrist flexors | 0 | 2 | 2 |
| Wrist extensors | 0 | 1 | 2 |
| IP flexors | 0 | 2+ | 3 |
| IP entensors | 0 | 1+ | 2 |
| Hip flexors | 0 | 3+ | 4- |
| Hip extensors | 0 | 2 | 2+ |
| Hip abductor | 0 | 3+ | 4 |
| Knee extensors | 0 | 4+ | 4+ |
| Knee flexors | 0 | 3 | 3 |
| Ankle dorsiflexors | 0 | 2 | 3 |
| Ankle plantaflexors | 0 | 3+ | 4+ |

*Using Oxford muscle grading method

**Seven weeks post-admission:** The patient was re-assessed prior to discharge and the following were observed:

1. General bone alignment of the shoulder was maintained, hence, no subluxation.
2. Selective muscle power testing (table 1) showed remarkable improvement in muscle strength, especially in the left lower limb muscles.
3. Moderate reduction in elbow and knee flexor spasticity. Grade on Bronstom assessment was 4 (see appendix). On admission the grade was zero.
4. She could now ambulate without the back slab, although with typical hemiplegic gait.
5. Remarkable reduction in facial deviation.
6. General functional activity in the hand was still reduced as she could grip but did not have the power to open fingers and release.
7. Remarkable reduction in shoulder, knee and ankle joint pains.
8. Ankle clonus had stopped.

The management programme was maintained at three sessions per week for another 4 weeks on outpatient basis. Electrical stimulation of the interphalangeal (IP) joint extensors was commenced due to the poor response of the extensors to management using the previous mode of stimulation, cycle ergometry (for 5 minutes) and overhead reciprocal pulley exercises were included in the programme. Though the previous line of management continued, it was observed that the muscle groups previously stimulated now responded at a lower current intensity and voltage when compared to the initial stage.

**Four weeks post-discharge (that is 11 weeks post-admission):** Reassessment of the patient showed general improvement. The facial deviation had almost been totally corrected and there was remarkable reduction in the general spasticity initially exhibited by the patient. The gross muscle power had improved to 3 in the left upper limb muscles excluding the IP extensors that had a grading of 2. Also, the gross muscle power in the left lower limb had improved to 4 over the 11-week period of the rehabilitation programme. There was no muscle atrophy in the shoulder and arm muscles. An increment of 0.2 and 6.7 percent in muscle bulk was observed for the shoulder and arm girth respectively. At the time of this study, RB was on a lively splint for the IP joint extensors.

## DISCUSSION

A remarkable reduction in spasticity was recorded using either direct stimulation of the spastic muscle or the stimulation of the antagonist muscle in conjunction with other modalities. There was remarkable reduction in the spasticity exhibited by the left elbow flexors and the knee flexors following 11 weeks of electrical stimulation at the rate of three sessions a week. The reduction could not, however, be attributed to the use of electrical stimulation alone because other modalities were also used. There was no glenohumeral subluxation in the left shoulder joint and this could be attributed to the muscular

**Table 2.** Girth measurement

| | On admission | | 7 weeks post-admission | | 11 weeks post-admission | | Percentage increase in muscle bulk when compared to value on admission for left upper limb |
|---|---|---|---|---|---|---|---|
| | Right | Left | Right | Left | Right | Left | |
| Shoulder girth* | 21.8cm | 21.7cm | 22.0cm | 12.5cm | 22.0cm | 21.8cm | 0.5% increase |
| Arm girth** | 22.0cm | 21.0cm | 21.8cm | 19.5cm | 21.8cm | 20.8cm | 0.1% decrease (6.7% increase between the 7th and the 11th week) |
| Mid thigh girth*** | 45.6cm | 45.1cm | - | - | 45.5cm | 45.2cm | 0.2% increase |

Landmark:
*3cm from the acromion
**7cm from the acromion
***15cm from the left anterior superior spine

integrity of the shoulder muscles that were maintained using electrical muscle stimulation. Similarly, no obvious atrophy was noticed in all the groups of muscles that were stimulated (table 2) excluding a 0.1% decrease obtained for arm girth. A 6.7% increase was, however, observed for the arm girth between the 7th and 11th week. The results obtained in this case study support the findings of previous investigators[3, 4, 5] on the effect of electrical stimulation on spastic muscles and in the prevention of atrophy, because a very low percentage decrease (0.1%) was observed for the arm girth between the time of admission and the 7th week.

## CONCLUSION

The case study did not have a control, hence future experimental studies should include this. The rehabilitation of this patient showed satisfactory recovery. Presently, the patient has attained independence in most functional abilities; she is now able to climb stairs and is back in school. This case study has demonstrated that electrical stimulation, when used in conjunction with other physiotherapy modalities like infra red radiation, passive movement, proprioceptive neuromuscular facilitation, etc., is very effective in the management of hemiplegic patients.

## APPENDIX

**Bronstom Assessment**

0 - Complete flaccidity
1 - Onset of spasticity
2 - Remarkable spasticity
3 - Slight reduction in spasticity
4 - Remarkable reduction in spasticity
5 - Normal muscle tone

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