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A Case Report of Limb Dystonia Management in a Nigerian Hospital: Physiotherapy Perspective

Oti IK[1], Nwaedozie OC[2], Ayerite AB[3], Anyama EU[4]
[1,2,3,4] Neurology Unit, Department of Physiotherapy, University of Port Harcourt Teaching Hospital, Port Harcourt, Rivers State

ABSTRACT
Background: Dystonia is a common movement disorder characterized by abnormal postures of the affected body part. The disabilities associated with dystonia are socially disabling and disfiguring, ranging from pain, loss of muscle strength and activities of daily living, poor quality of sleep and even depression. This article presents a case report of physiotherapy approach in the management of a patient with limb dystonia with major focus on pain management, balance control and functional independence.
Case Presentation and Treatment Outcome: A case report of the management of a 48-year old female patient with limb dystonia. Patient was referred from the neurology unit, department of Internal Medicine, University of Port Harcourt Teaching Hospital (UPTH) to the neurology unit of the physiotherapy department, UPTH as a case of lower limb dystonia with an unknown cause. Appropriate physiotherapy intervention was given for a period of 12 weeks after a thorough assessment of the patient. The Numerical Pain Rating Scale (NPRS) was used to assess the level of pain in the affect limbs. The Berge Balance Scale (BBS) was used to assess the patient's balance while Barthel Index (BI) was used to assess the functional independence of the patient. The case report was presented in line with CAse REport (CARE) guideline for case reports. All ethical procedures were followed. Treatment Outcome revealed that the score on Berg Balance Scale increased from 25 to 45 while that of Barthel Index scale increased from 13 to 18 after 12 weeks of intervention. Pain level on Range of Motion (ROM) based on the Numerical Pain Rating Scale (NPRS) reduced from 4/10 to 0/10 on the upper limb and from 9/10 to 1/10 on the lower limbs.
Conclusion: Physiotherapy is effective in the management limb dystonia. Early referral to Physiotherapy is key to pain management, balance control and overall functional independence of the patients.

KEYWORDS: Limb dystonia management, Physiotherapy perspective, Nigerian Hospital, Case report

INTRODUCTION

Dystonia is a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal and often repetitive movements, postures, or both [1]. It is often initiated or worsened by voluntary action and is associated with overflow muscle activation [2]. Dystonic movements are typically patterned, twisting, and may be tremulous. Research has investigated the health-related quality of life (QoL) in patients with dystonia and indicated a poorer QoL in these patients [3]. Several causes affect QoL in patients, including impaired physical functioning owing to the presence of motor symptoms. The most severe, generalized forms of dystonia can affect all activities of daily living such as walking or hand function.

Dystonia is classified by age of onset, distribution of affected body parts and aetiology [4]. Dystonia can develop at any age, early onset (< 26 years) and late onset (> 26 years). Those with earlier age of onset are more likely to have a severe course affecting more of the body. Dystonia by distribution divides patients based on whether it is localized to a single body (focal), has spread to contiguous (segmental) or non-contiguous (multifocal) regions, or if the legs are affected along with other body regions (generalized). According to Albanese *et al* [4], Dystonia is best described by associated nervous system pathology, genetic or acquired causes or idiopathic (divided into sporadic and familial).

Patients with dystonia represent 20% of patients in movement disorder clinics [5]. Dystonia cases account for 15-30 cases per 100,000 in the general population [1]. In a study of a random sample of the population over 50 years of age, the prevalence of isolated dystonia was concluded to be 732 per 100 000, suggesting that in the ageing population dystonia is a common neurological disorder [5]. Women are more likely to be affected by this condition than men with a ratio of 2:1 [5]. Adult-onset focal dystonia syndromes are by far the most frequent presentations of this disease [4].

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The exact cause of dystonia is not known [6]. It can be caused by a problem with the part of the brain that controls movement. It can be hereditary or a symptom of another disease or condition, including: Parkinson's disease, Huntington's disease, Wilson's disease, traumatic brain injury, birth injury, stroke, brain tumor, oxygen deprivation or carbon monoxide poisoning, infections, such as tuberculosis or encephalitis and reactions to certain medications or heavy metal poisoning [7].

The symptoms of dystonia may be continuous or on and off [3]. They may be triggered by stress, fatigue or anxiety. They include: uncontrolled muscle cramps and spasms, parts of the body twisting into unusual positions such as the neck being twisted to the side or the feet turning inwards, shaking (tremors) and uncontrolled blinking. Complications of dystonia can be physical disabilities that affect the performance of daily activities or specific tasks, difficulty with vision that affects the eyelids, difficulty with jaw movement, swallowing or speech, pain and fatigue, due to constant contraction of the muscles, depression, anxiety and social withdrawal [7].

DIAGNOSIS AND MANAGEMENT OF DYSTONIA

In the diagnosis of dystonia, the clinical presentation will determine the appropriate test to be done. History of birth injury, family history of other neurological disorders or exposure to dystonia-inducing drugs is important [8]. MRI can be carried out as well as some blood and urine tests. Treatment of dystonia depends on the cause and type of dystonia. It involves a multidisciplinary approach which includes the use of medical, surgical, a combination of both and physiotherapy management [9].

Physiotherapy treatment is specific to the individual needs and aims to maximize patient's potentials, quality of life and independence. It includes: exercises to improve coordination and facilitate normal patterns of movement; exercises to reduce stiffness and prevent muscle contractures; exercises to strengthen weak muscles, postural exercises to improve symmetry and alignment, pain management techniques such as use of heat/ice to reduce pain associated with muscle spasm, relaxation techniques, positioning and appropriate seating, provision of splints to help control posture and reduce tone and hydrotherapy treatment to reduce muscle stiffness and aid relaxation.

OUTCOME MEASURES

The Berg Balance Scale (BBS) is used to objectively determine a patient's ability or inability to safely balance during a series of predetermined tasks. It is a 14 item list with each item consisting of a five-point ordinal scale ranging from 0 to 4, with 0 indicating the lowest level of function and 4, the highest level of function. It takes approximately 20 minutes to complete. A score of 50 indicates functional balance and a score of less than 45 indicates individuals that may be at greater risk of falling [10].

The Barthel Index (BI) consists of 10 common functional ADLs assessing for independence/dependence. Eight of the ten items represent activities related to personal care: feeding, bathing, grooming, dressing, bowels continent, bladder continent, toilet use and transfer (bed to chair and back); the remaining 2 are related to mobility and stairs. The index yields a total score out of 20, the higher the score, the greater the degree of functional independence [11].

The Numerical Pain rating Scale (NPRS) is a scale of 0 to 10. Number 0 means no pain while number 10 denotes maximum pain.

CASE PRESENTATION

A case of a 48-year old woman, married with three children, a business woman by profession was referred from the neurology unit of UPTH to the neurology unit of the physiotherapy department. She was being managed for lower limb dystonia with an unknown cause. Radiological report showed multilevel intervertebral disc degeneration from L1to L5, mild spinal canal stenosis of L4-L5 disc level.

The presenting complaint was inability to walk without support, involuntary muscle twitch of the lower limbs, pain in the left groin and inability to engage in Activities of Daily living (ADL). Patient is not a known hypertensive and was recently diagnosed of Type 2 Diabetes Mellitus (DM). On observation, patient was afebrile to touch, acyanosed, anicteric, neither pale nor dehydrated, and in no any obvious respiratory distress. The blood pressure, pulse rate and respiratory rates were all normal.

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Objective examination showed that the right upper limb was full but painful at the end range of joint Range of Motion bilaterally (ROM) (NPRS = 4/10), sensation, proprioception, and stereognosis were intact bilaterally. Muscle tone was normal; grip strength was good, gross muscle power was 4 on the left upper limited and 5 on the right upper limb. Back extension was limited and painful, left trunk flexion was limited and painful on the right side, muscle tone of the abdomen was hypotonic. Gross muscle power (GMP) of both lower limbs were 3; Passive ROM (PROM) was full and painful at all range while active ROM (AROM) was limited and painful from middle to end range of the movement in all directions bilaterally (NPRS = 9/10) . Spasticity was present (cogwheel), tendon Achilles tightness was mild on both lower limbs, ankle clonus, Babinski reflex, hamstring and adductor tightness were present on both lower limbs.

Functionally, patient could not walk without support, could not stand or walk for long with support, could not stand from sitting without support, did not have control in standing to sitting, could not turn in bed and could not get up from lying unsupported.

On assessment with berg balance scale, patient had a score of 25 on Berg Balance Scale and 13 on Barthel Index scale at baseline. After 6 treatment sessions, patient had better control in sitting to standing; patient could stand for a longer time and could take some steps without support. Berg balance score increased to 32 while barthel index score increased to 16. After 12 weeks of physiotherapy intervention, berg balance score increased to 45 and barthel index to 18. Pain level based on the Numerical Pain Rating Scale (NPRS) reduced from 4/10 to 0/10 on the upper limb and from 9/10 to 1/10 on the lower limbs.

Physiotherapy Management

The objectives of physiotherapy management were to relief pain, improve muscle tone, ROM of the joints, Activities of Daily living (ADL), strengthen weak muscles, enhance balance control and improve functional independence. The means of achieving these aims were cryotherapy to the lower limbs, sustained passive stretching, mobilization, and graded resisted active exercises to the lower limbs, trunk stabilization exercises, free active exercises/mild resisted active exercises to the upper limbs, balance, precision and coordination exercises, standing and walking re-education.

Patient was seen 2 times a week for an average of 50 minutes in each session for a period of 12 weeks. The treatment outcome of the patient is summarized on the tables below:

Table 1: Treatment outcome for the upper limbs

	On Assessment (Baseline)		After 12 weeks of intervention	
	Right	Left	Right	Left
Gross Muscle Power	5/5	4/5	5/5	5/5
Fine Motor movement (Precision test)	Intact	Intact	Impaired	Intact
Sensory Integrity (Light & Deep touch)	Intact	Intact	Intact	Intact
Stereognosis	Intact	Intact	Intact	Intact
Muscle tone	Normal	Normal	Normal	Normal
Muscle Bulk	Nil obvious atrophy	Nil obvious atrophy	Nil obvious atrophy	Nil obvious atrophy
Proprioception (Joint position sense)	Intact	Intact	Intact	Intact
Grip strength	Good	Fair	Good	Good
Joint ROM	AROM full but painful at end range NPRS (4/10)	AROM full and painful at end range NPRS (4/10)	AROM full and pain-free at all ranges NPRS (0/10)	AROM full and pain-free at all ranges NPRS (0/10)
Hand-to- nose coordination test	Good	Fair	Good	Good

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Table 2: Treatment outcome for the lower limbs

	On Assessment (Baseline) Right	Left	After 12 weeks of intervention Right	Left
Gross Muscle Power	3/5	2/5	5/5	4/5
Sensory Integrity (Light & Deep touch)	Intact	Intact	Intact	Intact
Muscle tone	Spastic (cogwheel)	Spastic (cogwheel)	Improved Tone	Normal
Muscle Bulk	Nil obvious atrophy	Nil obvious atrophy	Nil obvious atrophy	Nil obvious atrophy
Proprioception (Joint position sense)	Intact	Intact	Intact	Intact
Joint ROM	PROM full but painful at all range while AROM was limited and painful from middle to end range NPRS (9/10)	PROM full but painful at all range while AROM was limited and painful from middle to end range NPRS (9/10)	AROM full but mildly painful only at end range NPRS (1/10)	AROM full but mildly painful only at end range NPRS (1/10)
Barbinski Reflex	Present	Present	Present	Present
Ankle Clonicity	Present	Present	Absent	Mild
TA contracture	Present	Present	Absent	Mild
Heel-to-shin coordination test	Poor	Poor	Good	Fair

Table 3: Treatment outcome based on outcome measures

Outcome Measures		On Assessment (Baseline)	After 6 treatment sessions	After 12 weeks of Intervention
Pain Level (NPRS) on Joint ROM	Upper limbs	4/10	1/10	0/10
	Lower limbs	9/10	5/10	1/10
Berge Balance Scale		25	32	45
Barthel Index		13	16	18

SUMMARY OF CASE PRESENTATION AND TREATMENT OUTCOME

This is a case report of Physiotherapy management of a 48-year old female patient with limb dystonia. Patient was referred from the neurology unit, department of Internal Medicine, University of Port Harcourt Teaching Hospital (UPTH) to the neurology unit of the physiotherapy department, UPTH as a case of lower limb dystonia with an unknown cause. Appropriate

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DISCUSSION

This article presented a case report of the physiotherapy management of a 48 year-old female patient with limb dystonia. Patient was referred from the neurology unit of the University of Port Harcourt Teaching Hospital (UPTH) to the neurology unit of the physiotherapy department, UPTH as a case of lower limb dystonia with an unknown cause. Physiotherapy intervention was given for a period of 12 weeks after a thorough assessment of the patient.

Steeves *et al.* [5] and Marsden [12] posited that women are more likely to be affected by dystonia than men with a ratio of 2:1. This assertion is in tandem with this case report in which the patient is female. In the same vein, previous studies have revealed that adult-onset focal dystonia syndromes are by far the most frequent presentations of this disease [13, 14]. The 48 year-old patient in this case study also had an adult-onset dystonia syndrome. According to Fahn [15], dystonia is the most common movement disorder seen by neurologists after Parkinsonism. Limb, axial, and cranial voluntary muscles can all be affected by the disease. In this case report, it is the limbs of the patient that were mainly affected.

The treatment outcome after the period of intervention revealed conspicuous improvement on the muscle tone, strength, pain level, balance, and overall functional abilities of the patient as seen in the scores of the Numerical Pain Rating Scale, Berge Balance Scale, and Barthel Index etc. Few studies have described physiotherapy approaches to relief pain, improve strength and enhance functional independence in individuals with dystonia [16]. According to Bleton [17], the approach of the physiotherapist to the management of each form of dystonia is individual-based and has to be specific. There is not one single method but several strategies related to the different clinical forms [17]. The approach to the management of the patient as presented in this case report is individual based and specific to the needs of the patient, thereby corroborating the assertion of Bleton [17].

CONCLUSION

Physiotherapy is effective in the management limb dystonia. Early referral to Physiotherapy is key to pain management, improvement in muscle strength, balance, coordination and overall functional independence of the patients.

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ETHICAL CONSIDERATION

All ethical procedures were followed.

CONFLICT OF INTEREST

The authors declared no conflict of interest.

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