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The efficacy of balance training in a Churg-Strauss Syndrome patient: a case report.

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ABSTRACT

INTRODUCTION

Churg-Strauss Syndrome is a rare small-vessel vasculitis associated with asthma, blood and tissue eosinophilia. Peripheral neuropathy is reported in more than 50% of cases of Churg-Strauss Syndrome and it is often found related to motor and functional recovery issues.

The Sensory system plays a key role in balance control, and that is why patients with polyneuropathy experience balance impairment during stance and gait. Impairments in gait and balance are two of the main causes of the falls characterizing Churg-Strauss syndrome. Evidence suggests that exercise programs can effectively improve gait and balance and reduce falls and fall-related injuries.

The purpose of this paper is to show the importance of balance focused physiotherapy interventions to provide the best functional recovery to Churg-Strauss Syndrome patients. To date, the most recent literature does not provide us with evidence focusing on rehabilitation and physiotherapy in those patients.

CASE PRESENTATION

A 65-year-old male affected by Churg-Strauss Syndrome was referred to our inpatient rehabilitation ward one month after the onset of symptoms.

Balance and gait were assessed using Berg Balance Scale, Mini-BESTest together with Tinetti Mobility Test.

The rehabilitation protocol was designed to focus on balance and proprioception training and was performed for 2 months, 5 sessions of 90 minutes/week. After one month 15 minutes of balance training with visual biofeedback using Biodex Balance System SD were included in the balance training sessions.

RESULTS

we performed 4 testing sessions at four different times: baseline (T0), one month after the beginning of training (T1), 2 months later the beginning (T2) and 6 months follow-up (T3). Berg Balance Scale values moved from 8 at baseline assessment to 50 at follow up assessment. Score on the Mini-BESTest showed an increase from 2 points at T0 to 24 points at T3. Tinetti Mobility Score values improved from 11 points at T0 to 24 points at T3.

DISCUSSION

Scores on the Berg Balance Scale, Mini-BESTest and Tinetti Mobility Test showed significant improvements on all the 4 testing sessions.

CONCLUSIONS

it can be thus said that rehabilitation programs focusing on balance training constitute a reliable enhancement for functional outcomes in patients affected by Churg-Strauss Syndrome. Nonetheless, further investigation on physiotherapy strategies is needed.

Keywords: gait, Churg Strauss, physiotherapy, balance, EGPA.

INTRODUCTION

Churg-Strauss Syndrome, commonly known as Eosinophilic Granulomatosis with Polyangiitis (EGPA), is a rare small-vessel vasculitis associated with asthma, blood, and tissue eosinophilia [1][2]. EGPA is one of the less common vasculitis, the annual incidence is estimated to be 0.11 to 2.66 new cases per 1 million people and the prevalence is approximately 10.7 to 14 per 1 million adults [3].

Said syndrome was described in 1951 by Jacob Churg and Lotte Strauss. EGPA is associated with extrapulmonary conditions too, showing an increasing severity when heart, central nervous system, peripheral nervous system, gastrointestinal tract and kidneys are affected too [4]. Peripheral neuropathy is reported in more than 50% of cases of EGPA and it often results in major issues in motor and functional recovery in patients. Long term outcomes in patients showed a satisfying overall remission rate [5] and EGPA has a lower mortality rate compared to other systemic vasculitis [6].

Functional and motor recovery, as well as the improvement of the quality of life is pivotal for EGPA patients and physiotherapy should be a key part of the recovery process. The sensory system plays a key role in balance control, so patients with polyneuropathy experience balance impairment during stance and gait [7]. Altered gait and balance impairment are two of the main causes of falls [8]. Falls and fall-related injuries are common among older people and patients with cognitive or functional impairments. Falls cause significantly negative effects on the quality of life, functional status, and independence of the patients. Thus, preventing falls and related injuries is a major public health challenge worldwide [9].

Evidence suggests that exercise programs can effectively improve gait and balance in people affected by the Churg-Strauss syndrome and reduce falls and fall-related injuries [10]. To date, literature shows no published reports focusing on balance training and physiotherapy in EGPA patients. The following databases were searched: PubMed, Cochrane Library, Scopus, PEDro; the following search terms were used: "churg-strauss syndrome" AND physiotherapy; EGPA AND physiotherapy.

Studies that were not written in English or Italian language were not taken into consideration. In this study we analyze and observe the results of physiotherapy and balance training in an EGPA patient. The aim of this study is to show the crucial role played by balance focused rehabilitation to help EGPA patients to reach the best functional recovery.

CASE PRESENTATION

A 65-year-old male, physician, affected by EGPA was referred to our inpatient rehabilitation ward one month after the onset of symptoms. According to his medical history he was diagnosed with polyneuropathy 6 years before that moment. He was previously treated with immunoglobulins with a complete remission. He was then admitted to the emergency department with fever and diarrhea. Blood tests and electromyography showed leukocytosis, eosinophilia, and common peroneal nerve bilateral axonal damage. The patient was admitted to the neurology ward and was diagnosed with EGPA.

Clinical findings and assessment

When the patient was admitted to the rehabilitation ward, he was unable to switch from sitting to standing without help and to stand without support. The patient was positive to Romberg's test and his standing balance was compromised. Further physical examination revealed that power was slightly reduced in the right lower limb with F 3/5 hip abduction, hip extension, knee extension, ankle eversion and toes extension, F 2/5 plantar flexion, toes flexion and F 0/5 big toe extension. Power was reduced in the left lower limb with F 4/5 knee extension, F 3/5 hip abduction, dorsiflexion, eversion and toes flexion, F 2/5 plantar flexion and F 1/5 knee flexion. All assessments were performed using the Medical Research Council (MRC) Scale. (Table 1)

Tone was normal in all four limbs. Joint proprioception and tactile sensitivity were compromised in the right foot sole and reduced in the right-hand volar side (2 Points Discrimination Test Threshold 2PDTT 7,0 cm) and dorsal side (2PDTT 6,5 cm). The assessment of balance and gait was conducted by means of the Berg Balance Scale [11], the Balance Evaluation Systems Test (Mini-BESTest) [12] and the Tinetti Mobility Test [13].

Such assessment tools were chosen based on evidence concerning gait and balance assessment in patients with polyneuropathy and neurological impairments. All testing sessions were executed by the same experienced physiotherapist in the same clinical setting.

MRC SCALE STRENGTH ASSESSMENT	RIGHT SIDE	LEFT SIDE
Hip abduction	F 3/5	F 3/5
Hip extension	F 3/5	F 5/5
Knee extension	F 3/5	F 4/5
Knee flexion	F 5/5	F 1/5
Ankle dorsiflexion	F 5/5	F 3/5
Ankle plantar flexion	F 2/5	F 2/5
Ankle eversion	F 3/5	F 3/5
Toes extension	F 3/5	F 5/5
Toes flexion	F 2/5	F 3/5
Big Toe Extension	F 0/5	F 5/5

Table 1: lower limb strength assessment using the MRC Scale

Treatment

Nevertheless, the balance and proprioception training were considered the main rehabilitation focus [14][15]. The rehabilitation protocol was performed for two months, five times per week, with 90 minutes session.

A typical training session included 10 minutes warm up and supervised walking, a 30 minute phase of strengthening and tactile sensibility training and 50 additional minutes of balance and proprioception training. After one month the balance training session was enriched with visual biofeedback using Biodex Balance System SD (BBSSD) [16].

The patient performed two blocks of 5 minutes of training with 5 minutes of rest in between. (Table 2) The decision to add the BBSSD training sessions is to improve balance abilities and to increase patient motivation, compliance, and commitment to the rehabilitation program.

Starting with supported standing balance exercises, more challenging exercises became possible, using supporting base reduction and single leg stance as instruments to improve the patient's performance. A standard lower limb muscle strength training was included in the program as well, in addition to a tactile stimulation focused on palms and soles.

TREATMENT WEEKS	COMPLETED TASKS
Week 1-2	<ul style="list-style-type: none"> static balance exercises with gradual upper limb support reduction static balance exercises without upper limb support and gradual supporting base reduction
Week 3-4	<ul style="list-style-type: none"> blinded static balance exercises and dynamic balance exercises
Week 5-6	<ul style="list-style-type: none"> balance training with visual biofeedback (Biodex Balance System SD) with upper limb support static balance exercises on unstable surfaces dynamic balance training with gradual supporting base reduction
Week 7-8	<ul style="list-style-type: none"> static single leg balance exercises with upper limb support static single leg balance exercises reducing upper limb support

Table 2: training program schedule

RESULTS

Rehabilitation program was performed for 2 months, 5 sessions of 90 minutes/week. All sessions were conducted without any complaints or any adverse event. No falls or other safety accidents occurred during the training sessions. The patient's treatment compliance, improvements and perceived discomfort were crucial in facilitating the rehabilitation process, as well as his ability to accurately report any adverse symptom. Four testing sessions were performed: the baseline (T0), one month after beginning of the training (T1), 2 months after the beginning of

the training (T2) and a 6-month follow-up (T3). Scores on the Berg Balance Scale, Mini-BESTest and Tinetti Mobility Test during the 4 testing sessions are shown in Table 3 and Figure 1.

	T0 (baseline assessment)	T1 (1 months assessment)	T2 (2 months assessment)	T3 (6 months assessment)
Berg Balance Scale	8	23	43	50
Mini-BESTest	2	4	16	24
Tinetti Mobility Test	11	17	19	24

Table 3: scores on the Berg Balance Scale, Mini-BESTest and Tinetti Mobility Test during the 4 testing sessions (baseline T0, one month T1, 3 months T2 and 6 months T3)

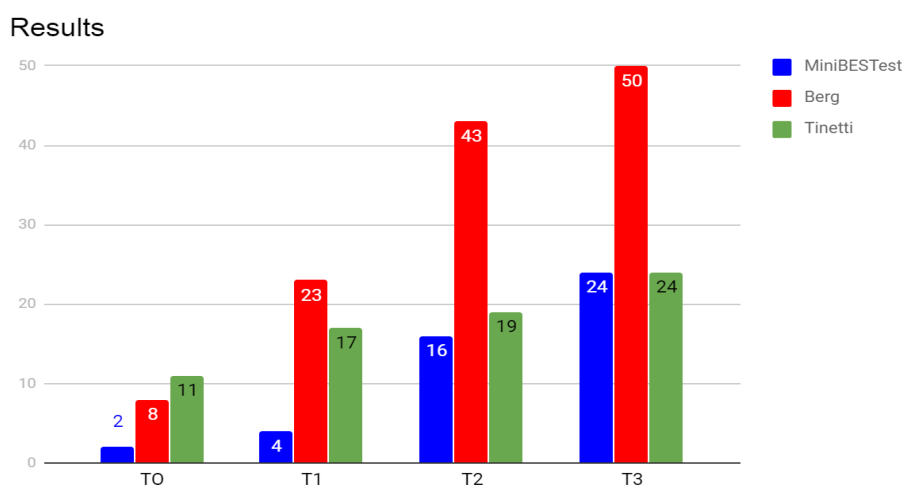


Figure 1: scores on assessment scales at 4 testing sessions

The scores obtained during the 4 testing sessions were registered on the Berg Balance Scale and resulted as listed below (see Table 4): at T0 8/56, at T1 23/56, at T2 43/56 and T3 50/56. Evidence shows that minimal detectable change (MDC) was identified from 2.8 to 6.6 points [13]. (Figure 2)

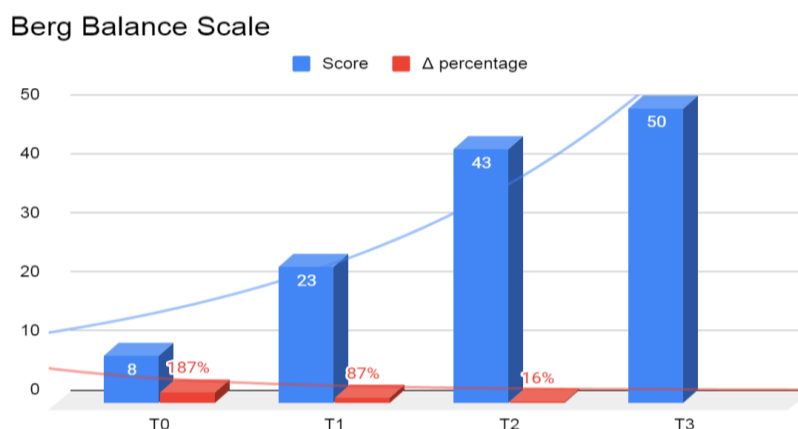


Figure 2: scores on the Berg Balance Scale

Figure 3 shows the scores which were registered on the Mini-BESTest these scores were recorded during the 4 testing sessions: T0 2/28, T1 4/28, T2 16/28 and T3 24/28.

MDC for Mini-BESTest was calculated in 3.5 points [12].

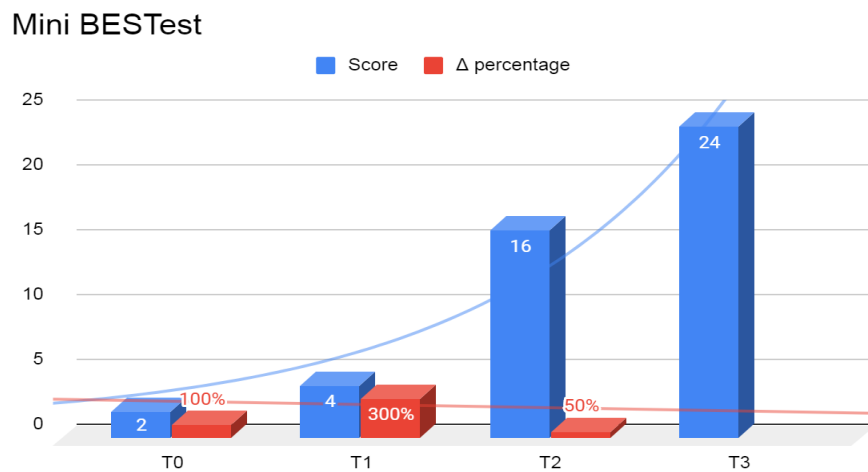


Figure 3: scores on the Mini BESTest

Figure 4 shows the scores which were registered on the Tinetti Mobility Score: T0 11/28, T1 17/28, T2 19/28 and T3 24/28.

Evidence shows that MDC for Tinetti Mobility Score is identified in 4 points in Huntington's Disease patients [17] and in 6 points in stroke patients [18].

Using a cutoff value of 21, the Tinetti Mobility Score has a sensitivity of 74% and a specificity of 60% to identify fallers [19].

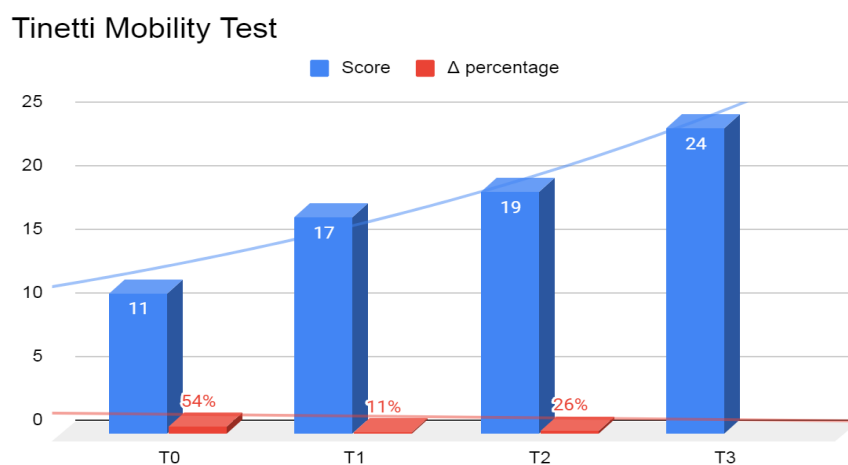


Figure 4: values on the Tinetti Mobility Score

DISCUSSION

Evidence shows that a targeted exercise program involving balance and functional exercises could crucially reduce falls [20]. An intensive program with daily physiotherapy sessions for two months was performed. After the first month, therapists decided to include a daily balance training with visual biofeedback by means of the Biodex Balance System SD.

The possibility to ensure visual feedback to the patient is to be considered a significant tool for therapists. It was very useful in improving the patient's commitment and adherence to the program as well. Substantial improvements were registered in all the balance and gait evaluation scales that were used to measure the patient's progress.

Berg Balance Scale showed a huge improvement from T0 to T1 and from T1 to T2 with changes higher than MDC; Scores improved dramatically from T0 testing session to T1 with 187.5% improvement scored. From T1 to T2 a 86.96% improvement was recorded and from T2 to T3 resulted in a further 16.28% improvement.

Higher enhancements than MDC were registered on Mini BESTest from T1 to T2 and from T2 to T3 too; values showed a 100% increase from T0 to T1 and a huge 300% increase from T1 to T2. An additional yet slight improvement of 50% from T2 to T3 was recorded at last.

Scores on the Tinetti Mobility Score are higher than MDC from T0 to T1 and from T2 to T3: it is in fact worth pointing out the 54% improvement from T0 to T1 and the small to mild improvements registered from T1 to T2 (11% increase) and from T2 to T3 (26% increase). Consistent enhancements regarding the patient's autonomy and everyday life activity management were registered too. As a consequence, the improvements occurred during the testing sessions, determined a crucial increase as far as the patient's self-belief and motivation is concerned. His abilities and autonomy kept growing during the whole rehabilitation program, allowing him to be able to be back home without the need of intensive assistance by the caregiver.

No safety issues or falls happened during the training sessions or during the rehabilitation period. This last result helps us explaining how prioritizing the role of balance training in preventing falls made the patient more confident about himself and his recovery chances.

The lack of a standardized assessment scales evaluating balance and gait validated on EGPA patients is to be considered a weakness of this study, and so is the absence of any specific rehabilitation program focusing on EGPA. The patient was not monitored after the 6 months follow-up, and this could also be a limitation to the present contribution.

CONCLUSIONS

Churg-Strauss Syndrome has a low mortality rate and shows good remission rate. Henceforth, it is physiotherapy and rehabilitation programs should be taken into consideration to allow pa-

tients to recover with as much autonomy as possible. It seems that rehabilitation programs focusing on balance training constitute a reliable way to improve functional outcomes in patients affected by EGPA.

To the authors knowledge, there is no scientific study taking physiotherapy for EGPA patients into consideration. Due to the lack of literature in this topic, this paper could be considered as a first step towards the definition of a shared and tailored rehabilitation and balance training program for EGPA. Further investigation on physiotherapy strategies is needed in order to provide the most suitable rehabilitation program for EGPA patients and stronger and more reliable evidence.

Learning points

- EGPA is often associated with peripheral polyneuropathy.
- Physiotherapy and balance training are very important in order to prevent falls and recover autonomy in patient with peripheral polyneuropathy.
- Including in the balance training program some visual biofeedback is very useful to improve patient treatment adherence and awareness of his condition.
- An intensive rehabilitation intervention should be considered as the most suitable option for EGPA patients with peripheral polyneuropathy.
- A well-structured rehabilitation program enhances the possibilities to return to a normal everyday life autonomy.

Patient perspective

At the beginning I felt really hopeless, and I thought it would be almost impossible to get over this situation and come back to my normal life.

During the rehabilitation program I was able to perceive my daily improvements and with the support of my family I started feeling more positive for my condition.

The support from my family and the rehabilitation team was crucial for my self-esteem as well. Now I feel safe and confident again walking around on my own and overcoming barriers.

My mood has clearly improved, and I feel positive about my future again.

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