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Rehabilitation Outcomes in a 75-Year-Old Male with Stiff Person Syndrome: A Case Study on Physiotherapeutic Interventions and Functional Improvement

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ABSTRACT: The following is a case for educational purposes involving a 75-year-old male, Parvez, with Stiff Person Syndrome (SPS) who has been referred to physiotherapy. The patient presented with balance and mobility concerns, increased rigidity and painful spasms in the lower extremities, and a mild plantar flexor contracture. Physiotherapy treatment included balance and gait training, strengthening and flexibility exercises, soft tissue release, and heat modality. Following treatment interventions, Parvez showed improvements in range of motion (ROM) and strength, as well as improved scores on the Activities-Specific Balance Confidence Scale (ABC scale), Rivermead mobility Index (RMI), Berg Balance Scale, Timed Up and Go (TUG), and Trunk Impairment Scale.

I. INTRODUCTION

Stiff Person Syndrome (SPS) is a rare neurological condition, diagnosed in approximately one in one million people^[1]. It is strongly related to autoimmune conditions (like diabetes type 1^[2]) and it affects the central nervous system and doesn't have any satisfactory treatment.^[3]The low occurrence of the condition may contribute to the limited research available regarding the effectiveness of a physiotherapy intervention. It is under diagnosed disorder which reduces the quality of life and the general mobility of the patients.^[2]

II. CLINICAL FEATURES

In individuals who have SPS, it is typical to present with a hyperlordotic posture, rigidity of truncal muscles and fluctuating muscle spasm^{[4] [5]}. The symptoms typically have an insidious onset with increased tone starting in the paraspinals, proximal lower extremities and abdominal muscles^{[4] [5]}.The **hallmark sign** of the disease is co contracture of agonist and antagonist muscle and continuous involuntary firing of motor units^[2]. Anticipatory anxiety and task specific phobias due to abrupt startle responses is also seen. Individuals with SPS may also present with episodic spasms particularly in the lower limbs that may have an associated trigger^{[4][5]}. The spasms may cause the patient to fall with a tinman-like posture^[5].Other neurological symptoms are gaze palsy, nystagmus, increased reflexes and paroxysmal dysautonomic crisis.^[3]

Differential Diagnosis

Multiple sclerosis

Parkinson's disease

Psychiatric disorders secondary to phobias and emotional triggers^[4].

III. REVIEW OF LITERATURE

In 2011, Hegyi reported a case concerning a 24-year-old woman who had been diagnosed with SPS for approximately one year. In Hegyi's report, the patient attended 17 sessions across 15 weeks of outpatient physio. The patient's chief concerns were pain, muscle spasms, gait anomalies and Range Of Motion (ROM) deficits particularly in the lower left extremities. In this case, the patient's physiotherapy interventions were designed to treat these issues. Treatment included: Therapeutic ultrasound, soft tissue mobilizations, manual stretching, therapeutic exercise, and fitting for an ankle-foot orthosis. These interventions were successful and improved the symptoms presented.

In another case reported by Potter in 2006, a 33-year-old man was being treated via in-patient physiotherapy for 10 days. He had been diagnosed with SPS for three years prior to the recorded physiotherapy intervention. The man presented with decreased static and dynamic balance, unable to maintain a standing position for more than



five seconds without handheld supports, and requiring use of a rollator walker to ambulate short distances^[5]. The man typically used a wheelchair in any attempt to ambulate. The patient presented with axial rigidity, hyperlordosis, and bilateral lower extremity rigidity^[5]. In examining his past medical history, it was apparent that he had suffered from anxiety, depression, substance abuse and chronic bronchitis. He also had pain that ranged from a four out of 10 when sitting quietly, which could at times reach 10 out of 10, depending on the circumstances.

The patient averaged 45 minutes a day of physiotherapy for 10 days, with treatment focused on improving functional independence and basic motor skills. Physiotherapy consisted of therapeutic exercises and functional retraining. The patient was given a home exercise program that focused on stretching his lower extremities and flexion-based truncal exercises. Although the physiotherapy treatment was scheduled for a longer period, the patient requested early discharge and even with reduced treatment time, his function improved. In this case, the patient appears to have a much more advanced instance of SPS compared to the fictional patient case we will explore, however the goals and physiotherapy management remain similar. I can use the case of Parvez to support the efficacy of the physiotherapy for a patient with SPS which has been questioned in the past^[4].

In the following fictional case, my patients name is "Parvez". Parvez is a 75-year-old man who was diagnosed with classical SPS approximately one year ago. Parvez presents increased axial stiffness, and bilateral lower extremity stiffness, with his right side more affected compared to his left lower extremity. Parvez presents with hyperlordosis, low back pain, hypertonic trunk musculature and hypertonic lower extremity musculature. Although SPS is a rare neurological disorder and there is limited evidence regarding the efficacy of physiotherapy as a treatment, the two cases above support the indication of physiotherapy for Parvez's case. The objective of this case study is to provide more information and literature regarding the efficacy of physiotherapy applied to an individual with SPS in an outpatient setting.

IV. Patient Characteristics

Parvez (further known as Mr. P) is a 75-year-old male. He is a retired data analyst, who lives at home with his wife in Surat. Mr. P had noticed decreased ability to balance, beginning two and a half years ago, leading to increased incidents of falling (his last fall being around two weeks ago) which had progressively gotten worse, and low back pain, starting a year and a half ago. Within the last year he noticed increased pain in his lower extremities accompanied by muscle spasms, particularly affecting the left leg. He was referred to a neurologist and diagnosed with SPS. He received a referral for outpatient physiotherapy to perform a falls risk assessment, help with his balance and gait, manage muscle rigidity and spasms and maintain his independence and quality of life.

V. EXAMINATION FINDINGS

Subjective

- **Patient Profile (PP):** 75 y/o male, right hand dominant
- **History of Present Illness (HPI):** Diagnosed with Stiff Person Syndrome 1 year ago, left plantar flexor contracture (~2 months), muscle spasms, and progressive rigidity (~1 year) and lower extremity pain (~6 months), chronic low back pain (~1.5 years).
- **Past medical history:** Type 1 Diabetes, hypertension, high cholesterol.
- **Medications:** Oral diazepam, gabapentin^[6], and insulin
- **Health Habits:** Non-smoker, drinks occasionally (~2 drinks a week).
- **Psychosocial:** The patient describes feeling lonely due to COVID-19 as well as frustrated with the diagnosis. Over the past 6 months he has avoided gardening and groceries due to feeling unsteady and a fear of falling. He lives with his wife; his son lives 20 minutes away and visits weekly; his daughter lives 1 hour away and visits monthly.
- **Home:** Bungalow, 5 stairs into home with railings on the right at both front and back door. The bathroom has a stand-up shower with railings.
- **Previous Functional status:** Able to ambulate more than 200m with no gait aid, active in gardening and golfing, no issues performing activities of daily living (ADLs).
- **Current Functional status:** Less confident walking outdoors, feeling unsteady, rigidity and painful spasms in both lower extremities prevented him from engaging in his hobbies (gardening, watching golf, sailing).



- **Imaging/diagnostic testing:** MRI and X-ray for chronic low back pain (all negative), Blood tests (Anti-GAD body 92.5 units/mL), nerve conduction studies (No abnormalities).
- **Precautions/contraindications:** None
- **Chief complaint:** decreased balance control, increased rigidity, and painful spasms in lower extremities particularly in the left leg limiting his ability to do ADLs.

VI. OBJECTIVE

General

- Vital signs
- Pulse 80bpm, BP 145/95mm HG, RR 15 bpm
- Pain
- Visual analogue scale
- 3/10 constant pain in low back
- 4/10 during painful spasms in the right lower extremity, caused by hot weather and stress
- 7/10 during painful spasms in left lower extremities, caused by hot weather and stress

Posture

- Thoracic kyphosis
- Hyperlordosis

Gait

- Slow and wide gait, appearance of stiff-legged walking
- Left ankle is stiff and lacks dorsiflexion during gait
- Max walking distance 200m
- Typically ambulates with a 4WW outdoors and single cane indoors
- Shuffling
- Decreased arm swing

Tone

- Increased tone in the lower extremities, more increased tone on the left
- Mild plantar flexion contracture, as well as lack of knee extension

AROM

- Knee flexion (right 135°, left 120°)
- Knee extension (right -10°, left -20°)
- Ankle dorsiflexion (10° right, left -5°)
- Ankle eversion (10° right, left 5°)
- All other ROM within normal limit (WNL)

PROM

- Knee extension (right -5°, left -10°)
- All other ROM within normal limit (WNL)

Strength

- Right lower extremity (4/5)
- Left lower extremity (3+/5)

Sensation

- Intact upper and lower extremities

Neurological testing^[7]

- Myotomes:4+ for all lower extremities, upper extremities normal
- Dermatomes:(normal)



- Reflexes: Hyperreflexia

Self-Reported Outcome Measures^[8]

- Activities-Specific Balance Confidence Scale(ABC)
 - score: 50% (moderate level of physical functioning)
- Rivermead Mobility Index (RMI)
 - 7/15

Outcome Measures ^{[5][8]}

- Berg Balance Scale (BBS)
 - 40/56
- Timed Up and Go (TUG)
 - 30 seconds
 - 0.5m/s gait speed
- Trunk Impairment Scale
 - 10/23

VII. CLINICAL IMPRESSIONS

The patient is a 75-year-old male with mild SPS. His subjective interview indicates that in his initial diagnosis he was still able to participate in his regular ADLs, but for the last six months has avoided doing his regular ADLs due to feeling unsteady, having progressive rigidity, and a fear of falling. Major clinical findings from the objective assessment revealed hyperlordosis, mild left plantar flexor contracture and lack of ROM in his ankles and knees, with his left ankle dorsiflexion and knee extension particularly affected. He also presents with chronic low back pain, rigidity, painful muscular spasms in lower extremities (left affected more than right), and decreased trunk and lower extremity strength.

His self-reported ABC score was 50%, indicating that he is moderately confident with his balance and has a fear of falling^{[9][10]}. This shows that any lack of participation in his ADLs is mostly due to his lack of confidence and fall risk^{[9][10]}.

The patient's lower extremity and trunk strength likely plays a role in impeding his balance. His trunk impairment score was 10/23, indicating that his sitting static and dynamic balance is poor^{[8][11]}. His BBS score is 40, which signifies that he is at an increased risk for falls (<45 indicates increased risk of falls)^{[12][13]}. His TUG score is in the 30s indicating that he is at an increased risk of falling (normal for his age is 9s+/-3s)^{[14][15][16]}. His RMI score is 7/15; a higher score would demonstrate better mobility performance^{[17][18]}. These findings highlight a need to include balance, gait intervention, and reassessment of appropriate gait aids in his treatment plan in response to his overall increased risk of falls.

Mr. P's health prior to the diagnosis of SPS was fair; he had type 1 diabetes, hypertension, and high cholesterol. However, he did partake in some activities and was relatively active for his age. His activities have been limited for six months now by his fear of falling, progressive rigidity, and painful spasms in the lower extremities, with his left side being worse than his right. It is believed that Mr. P would be a good candidate for physiotherapy to assist with improving his confidence, balance, and strength while reducing his muscle rigidity. Furthermore, Mr. P's care should extend to a multidisciplinary approach which will be explored later in this case.

Problem List

1. Decreased balance
2. Decreased confidence and increased fear of falling
3. Hypertension
4. High cholesterol
5. Type 1 diabetes
6. Lower back pain
7. Lower extremity pain (due to muscular spasms)
8. Decreased left ankle dorsiflexion and eversion ROM
9. Mild ankle plantar flexor contracture
10. Decreased left knee flexion and knee extension ROM



11. Kyphotic posture in thoracic spine
12. Hyperlordosis in lumbar spine
13. Decreased lower extremity strength
14. Rigidity
15. Hypertonia
16. High Anti GAD levels
17. Abnormal gait

Short term goals

1. Improve his ABC score from 50% to 64% in 6 weeks
2. Decrease his lower back pain from a 3/10 to a 1/10 at rest in 5 weeks
3. Decrease his pain from muscle spasms from a 7/10 to a 4/10 in the left leg and a 4/10 to a 2/10 in the right leg, in 5 weeks
4. Increase his left ankle dorsiflexion ROM from -5° to 0° in 6 weeks
5. Increase his left ankle eversion ROM from 5° to 8° in 6 weeks
6. Increase his knee extension AROM from -10° to -5° on the right and -20° to -15° on the left in 5 weeks
7. Increase his left knee flexion ROM from 120° to 127° in 6 weeks
8. Improve his lower extremity strength from a 4/5 to a 5/5 on the right and a 3+/5 to a 4/5 on the left side in 6 weeks
9. Improve his TUG score from 30 to 18 in 6 weeks
10. Improve his trunk impairment score from a 10 to 15 in 6 weeks

Long term goals

1. Improve his BBS from 40 to 47 in 16 weeks
2. Improve his RMI score from 7 to 12 in 16 weeks
3. Walk around the block with his wife with improved gait mechanics in 16 weeks
4. Garden for 30 minutes a day in 16 weeks

Treatment Plan

	Frequency	Intensity	Time	Rationale
Education <ul style="list-style-type: none"> • Role of PT • Pain management • Environmental modifications • Appropriate use of gait aids 	During his initial appointment and then in subsequent check-ins	N/A	Ongoing	Problem List 6, 7, 17 To make sure the patient understands the role of the physiotherapist, how to manage his symptoms and what needs to be done in order to see improvements
Balance training ^[19] <ul style="list-style-type: none"> • Standing shoulder width apart <ul style="list-style-type: none"> ○ Eyes open ○ Eyes closed ○ Foam surface • Standing tandem stance • Stand heel toe • Picking up objects off the floor • Perturbations 	In clinic: 2x/week for the first 8 weeks then progressing to 1x/week At home: 2x/week at home for the first 8 weeks then progressing to 3x a week at home	As tolerated, but actively trying to push past limits	Initially 15 minutes a day then progressing to 30 minutes	Problem list 1, 2 Increase Mr. P's confidence with his balance and decrease his risk of falls.



- Forewards
- Backwards
- Sideways
- Turning around (360s) in parallel bars

Gait ^{[5][20]}	In clinic: 2x/week then progressing to 1x/week	Light intensity	10 minutes initially progressing to 15 minutes	Problem list 1, 2, 17 Improve Mr. P's gait mechanics, walking endurance as well as increase his confidence and decrease his risk of falls.
<ul style="list-style-type: none"> ● Walk with nordic poles ● Parallel bar walking ○ Forewards ○ Backwards ○ Sideways ○ Over obstacles 	At home: walking with nordic poles 3x a week then progressing to 4x/week at home			

Strengthening ^{[5][4][21]}	At home:3x/week In clinic:2x/week then progressing to 1x/week(sit to stand, step ups, and kneeling to standing)	65%of 1RM/light intensity	3 sets of 12 reps	Problem list 11, 12, 13, 15 Postural strengthening and motor control are a key portion of both gait and balance, therefore taking steps to improve Mr.P's posture/motor control will help improve his gait and balance. Furthermore, strengthening of the lower extremity will lead to improvements in balance and gaits, which will allow Mr. P to return to his ADLs at a quicker pace.
<ul style="list-style-type: none"> ● Kitchen sink exercises ● Isometrics ○ Quadriceps ○ Calves ○ Hamstring ● Postural strengthening/motor control ● Sit to stand ● Step-ups in parallel bars ● Kneeling to standing and vice versa 		Light intensity	2 sets of 15 reps	

ROM ^{[5][4]}	At home:3x/ week progressing to 6x/week	Light pressure to start then proceeding moderately.	30s static stretch repeated twice a day	Problem list 8, 9, 10, 14, 15 Improvement of his ROM will translate to better gait and balance and will speed up his ability to participate in his ADLs.
<ul style="list-style-type: none"> ● Trunk ● Hip ● Knee ○ Extension ○ Flexion ● Ankle ○ Dorsiflexion ● General ROM with stationary bike 	In clinic:2x/week then moving to 1x/week(stationary bike)	Stretching to the point of slight discomfort until a stretching sensation is felt.	For 5-10 minutes before ROM exercises	
		Light intensity		

Additional treatments ^{[4][22]}	In clinic: 2x/week moving to 1x/week	5-10 Passes	Ongoing	Problem list 8, 9, 10, 14, 15 Improve his muscle extensibility and joint mobility allowing for
<ul style="list-style-type: none"> ● Soft tissue mobilizations of lower back and lower extremity muscles ● Joint mobilizations of 		As tolerated	10 minutes 3	



lower extremity joints
• Heat therapy as needed at home times a week improvements in Mr. P's overall mobility.

NOTE:

- The following exercises were not completed in one session; they were used throughout the 16-week treatment session as the patient progressed.
- These movements should be performed with caution in order to prevent muscle spasms.^{[23][24]}
- Interprofessional Health Team^[25]

It is important to include a variety of healthcare professionals in Mr. P's care plan. For instance, his family physician plays a crucial role in managing his acute and chronic conditions and determining other underlying conditions he may have^[26]. As well, family physicians are one of the first point of contact in the healthcare system and can refer to other specialists when needed. Neurologists are also vital in Mr. P's care as they are specialized to diagnose, treat, or manage Stiff Person Syndrome. They could prescribe the appropriate medication to manage his symptoms, perform tests such as nerve conduction studies and electromyography to evaluate the integrity of the nervous system as the condition progresses, and provide appropriate interventions to manage the condition when needed^[27].

In addition to his family physician and neurologist, there are other professionals that need to be involved. Considering Mr. P's age, high-cholesterol, and type 1 diabetes, it was viewed as essential that he was referred to a dietitian. Considering Mr. P's comorbidities, and complexity as a case, a professional opinion was required. A dietitian can provide vital advice to assist Mr. P with his self-management of his diabetes and high cholesterol which can assist in reducing long term complications commonly involved with diabetes and improve his quality of life^[28].

A social worker may also be included in his plan of care, as they can provide resources and services to overcome barriers with regards to aspects such as psychosocial, physical, and emotional well-being^{[29][30]}. In Mr. P's case, a social worker may direct him to appropriate resources as he reported feeling socially isolated due to COVID and frustrated with his diagnosis. A social worker may also help implement interventions to prevent caregiver burden that his wife may experience if Mr. P's condition was to progress.

As well, an occupational therapist may be included to help enhance Mr. P's quality of life and become more independent with his ADLs. They can provide assessments to determine whether adaptive equipment or home modifications are needed as his physical ability changes^[31]. Furthermore, they can determine if appropriate adaptations should be made or if new skills are to be learned, to allow Mr. P to engage in his gardening hobby, go grocery shopping, or perform his ADLs with ease^[31]. Occupational therapists could also provide education and guidance to Mr. P's spouse about how to provide accommodations and care when needed^[31].

VIII. OUTCOME

Based on the initial assessment, it was determined that Mr. P should receive four months of physiotherapy to address his impairments and implement a home exercise program. He was seen twice a week for the first eight weeks, and once a week for the remaining eight weeks. He was reassessed at the end of his six month treatment to determine how to proceed with ongoing therapy.

Major outcomes included: Increased ROM, improved overall balance and confidence, balance control, tone, gait mechanisms and pain reduction/management. While he now feels more confident gardening and walking around the block, he still has mild balance impairments, thoracic kyphosis, rigidity, and mild mobility concerns. His current scores are:

- ABC: 70%
- RMI: 10/15
- BBS: 47
- TUG: 13s
- Trunk Impairment Score: 18/23



After 16 weeks of the prescribed treatment Mr. P's BBS score improved to a 47/56 which indicates a minimal detectable change occurred and along with a decreased risk of falls^[12]. His ABS score improved from 50% to 70%, which still leaves him in the moderate level of physical functioning category. However, this change indicates a minimal important difference (range 11-13%) and that he is no longer at a fall risk as his score is above a 67%^{[9][10]}. His TUG score improved to 13s, which further underscores a lower risk of falls and more functional independence^{[14][15][16]}, and his RMI score improved to 10/15, indicating better mobility performance^{[17][18]}.

Overall, Mr. P achieved all his short-term and most of his long-term goals during the 16 weeks of treatment. Mr. P's RMI score improved significantly but did not meet his long term goal number two. It was decided at the end of the 16 weeks that the frequency of his visits would decrease to once a month, and additional long-term and short-term goals were set. A one-month follow-up was scheduled to check-in, reassess, and provide progressions to his home exercise treatment plan; this is a progressive disorder and the focus remains on maintenance and continued communication between Mr. P's interprofessional health team.

IX. DISCUSSION

Mr. P (75 y/o) presented with mild Stiff Person Syndrome. His initial assessment showed he had hyperlordosis, mild left plantar flexor contracture, decreased ROM in his ankles and knees (particularly in the left), rigidity and painful muscular spasms in the lower extremities (left being more affected). Our assessment also showed he had chronic low back pain and decreased trunk and lower extremity strength. Mr. P was independent in his ADLs but was becoming more dependent as he was avoiding physical activity due to his rigidity, muscle spasms and fear of falling. Therefore, his treatment plan was aimed to first decrease his rigidity while increasing his balance control, lower extremity ROM, strength, gait mechanics and balance confidence. Treatment would then progress to maintaining function, as SPS is a progressive disorder and the goal is ultimately to ensure Mr. P is able to continue performing his ADLs and hobbies independently.

Mr. P's treatment plan was specifically related to his goals and diagnosis of SPS, however due to the rare nature of SPS not much is known with regards to best treatment approaches, especially since individuals present differently. Therefore, our treatment plan was based on previous case studies^{[4][5]} his goals, chief complaints, assessment findings, as well as personal experience from treating an individual with SPS. Mr. P's primary concerns were his increased falling tendency, rigidity, painful spasms in the lower extremities and lack of confidence in his balance. As such, most of our treatment plan focused directly on improving his balance and gait, and increasing his lower extremity strength and ROM to assist in preventing future falls. Aspects of the treatment plan were modelled off the BBS (such as turning 360 degrees and picking items off the floor) because Mr. P had set goals to improve various outcome measure scores. Additionally, there was a focus on strengthening and stretching Mr. P's trunk and bilateral lower extremities to reduce rigidity, spasms and help work towards his goal of walking with his wife.

One aspect of treatment linked to his goals was having him perform kneeling to standing movements, and vice versa. Kneeling to standing not only works on strength and balance but it also allows Mr. P greater function with respect to many aspects of gardening (one of his hobbies)^[32]. Additionally, the ability to go from kneeling to standing helps Mr. P feel more confident in getting back up if a fall should occur, establishing greater confidence in participating in his hobbies and other physical activity^{[33][32]}.

The outcome measures chosen for Mr. P were based on previous case studies^{[5][4][8]} as well as personal experience. All the outcome measures chosen in this case study are not specific to SPS, however, most of them have validity in the older adult population. The exception is for the trunk impairment score, which was created and validated for stroke patients. It was selected as a measure for Mr. P based on other SPS reported case studies^[8], and allowed the ability to monitor and compare changes to his trunk control^{[8][11]} from pre to post treatment. Monitoring changes in trunk control was an important aspect for treatment as trunk control plays a significant role in balance and postural control^{[34][35]}— one area that Parvez had problems with. RMI is the other exception chosen for Mr. P as it was based on other reported SPS case studies^[8] but, created and validated for stroke patients. The RMI allowed for a self-report measure on Mr. P's mobility with tasks related to balance and gait, as well as other more functional activities^{[17][18]}.



Throughout the case study, it was determined that physiotherapy management of Mr. P's balance, gait, posture and pain was effective. It must be noted that the patient's highly motivated attitude towards physiotherapy may have contributed to this outcome. Another factor that contributed to the patient's success with physiotherapy was that his condition was considered to have been mild at the time of diagnosis. Considering the outcome it is clear that physiotherapy should be applied to individuals with SPS, as it can assist in managing and improving their symptoms^{[4] [5]}.

Overall, this case attempted to showcase a treatment path for an individual with mild SPS. Each SPS patient may present with varying symptoms^{[4] [5]} and therefore require a different approach to treatment, personalized to effectively manage their condition. This case study is particularly unique in that the average age of SPS diagnosis typically occurs around the age of 40^[4]. At age 75, Mr. P had only been officially diagnosed for one year, making his situation uncommon compared to the other case studies looking at much younger individuals with a similar diagnosis. At Mr. P's age, there must be more considered during the physiotherapy management of his treatment. Considering his age, diagnosis, and commodities, if Mr. P were to have anything but a mild case of SPS, the prognosis may not have been as positive. It should be noted that with the progressive nature of the disorder, physiotherapy throughout the case was not a cure, but meant to improve and maintain Mr. P's functionality and independence. As research progresses in this area, it is possible that other treatments and outcome measures may be more appropriate and applicable to the described patient.

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