

Effect of home-based pulmonary rehabilitation in patients with idiopathic pulmonary fibrosis

Effetti della riabilitazione respiratoria domiciliare in pazienti con fibrosi polmonare idiopatica

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ABSTRACT

Objective: The aim of this study was to investigate the effects of a home-based pulmonary rehabilitation program on the functional outcome parameters in patients with idiopathic pulmonary fibrosis (IPF).

Design: A prospective study.

Patients: Seventeen patients diagnosed with IPF.

Methods: A home-based pulmonary rehabilitation program was carried out in 17 IPF patients for 12 weeks. Dyspnea severity during daily life activities (Medical Research Council Scale), pulmonary function (pulmonary function test), exercise capacity (6-minute walking test, 6MWD), and general health related quality of life (Medical Outcomes Short Form-36) were evaluated.

Results: A significant decrease in perceived dyspnea ($p = 0.003$) and leg fatigue ($p < 0.05$) severities, and an increase in the 6MWD ($p = 0.04$) and general health related quality of life scores (health perception, physical role, and emotional status subscores) were found after the program ($p < 0.05$).

Conclusion: Home-based pulmonary rehabilitation may reduce dyspnea and fatigue severities, and improve exercise capacity and health-related quality of life in patients with IPF. In the treatment of IPF patients, home-based pulmonary rehabilitation programs should be placed alongside the routine treatment options.

Keywords: Exercise capacity, home-based pulmonary rehabilitation, idiopathic pulmonary fibrosis, quality of life.

RIASSUNTO

Obiettivo: Scopo di questo studio era valutare gli effetti di un programma di riabilitazione respiratoria domiciliare sugli indicatori funzionali nei pazienti con fibrosi polmonare idiopatica (IPF).

Disegno dello studio: Studio prospettico.

Pazienti: Diciassette pazienti con diagnosi di IPF.

Metodi: Un programma di riabilitazione respiratoria della durata di 12 settimane è stato applicato in 17 pazienti con IPF. Sono state oggetto di valutazione l'intensità della dispnea durante le attività quotidiane (Medical Research Council Scale), la funzionalità respiratoria (prove di funzionalità respiratoria), la capacità di esercizio fisico (test del cammino di 6 minuti - 6MWD), ed il livello complessivo di qualità della vita legato alla salute (Medical Outcomes Short Form-36).

Risultati: Al termine del programma sono stati rilevati una riduzione significativa della gravità della dispnea percepita ($p = 0,003$) e dell'affaticabilità degli arti inferiori ($p < 0,05$), un incremento del 6MWD ($p = 0,04$) e punteggi del questionario sulla qualità di vita (nei sottogruppi di percezione del proprio stato di salute, ruolo fisico e di condizione emotiva) ($p < 0,05$).

Conclusioni: Un programma di riabilitazione respiratoria domiciliare può ridurre l'intensità della dispnea e dell'affaticabilità, migliorare la capacità di esercizio fisico e la qualità di vita relativa alla salute in pazienti con IPF. Nel trattamento dei pazienti con IPF i programmi di riabilitazione respiratoria domiciliari dovrebbero perciò essere inclusi nelle opzioni terapeutiche di routine.

Parole chiave: Capacità di esercizio, fibrosi polmonare idiopatica, qualità di vita, riabilitazione respiratoria domiciliare.

INTRODUCTION

Idiopathic pulmonary fibrosis (IPF) is a chronic progressive lung disease causing dyspnea, fatigue, dry cough symptoms and reduced exercise capacity

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and health related quality of life. Exercise capacity is limited due to impaired respiratory mechanics, ventilatory insufficiency, hyperinflation and musculoskeletal dysfunction in obstructive lung diseases whereas the most important factor limiting exercise capacity in IPF is circulatory impairment that causes exercise induced gas exchange impairment [1,2]. The exercise induced hypoxemia leads to inactivity during daily life with the disease progression, causes functional limitations in addition to pathophysiological impairments and decreases health related quality of life in IPF patients [3,4].

The positive effects of pulmonary rehabilitation programs on functional outcome parameters such as dyspnea, exercise capacity, and health related quality of life have been investigated and proven mostly in patients with chronic obstructive lung diseases. Pulmonary rehabilitation has also been accepted among the non-pharmacological treatment options as a means to reduce dyspnea severity, improve exercise capacity, and quality of life of IPF patients in the previous studies. However, although the positive effects of pulmonary rehabilitation programs in these studies have been reported, a standard pulmonary rehabilitation program for patients with IPF has not been developed yet. These programs are mostly out-patient-based, and durations and contents are not similar. In addition, study groups are small, outcome parameters are different and IPF is not distinguished from other restrictive lung diseases. Therefore it has been suggested in the previous literature that standard/optimal exercise-based pulmonary rehabilitation programs should be designed and their effects should be investigated for IPF patients [5-10]. Also, it has been accepted that exercise programs should be adapted to the patient's daily life so as to maintain the gained positive effects of the pulmonary rehabilitation program [11]. Home-based programs are a suitable option for pulmonary rehabilitation during daily life. In addition, these programs are effective, useful, simple, cheap, practical, and equivalent as an alternative to in/out-patient programs [12,13]. Thus, we aimed to investigate the effects of a home-based pulmonary rehabilitation program on the functional outcome parameters in patients with IPF.

METHODS

Patients

We enrolled IPF patients diagnosed with IPF according to the diagnostic criteria of the American Thoracic Society (ATS)/European Respiratory Society (ERS) consensus statement were enrolled [2]. Inclusion criteria were as follows: a) being clinically stable; b) treatment with no more than 20 mg of prednisone per day; c) not having any pulmonary infections such as pneumonia at least in the last 6 weeks; d) not having serious uncontrolled cardiovascular, psychological problems; e) not receiving supplementary oxygen therapy; f) having no neurological, inner ear, or orthopedic disease; g) being able to ambulate without assistance or assistive

devices; h) willingness to participate in this study. The exclusion criteria included the presence of any of the following: a) obstructive lung disease ($FEV_1/FVC < 80\%$) such as chronic obstructive pulmonary disease (COPD) or asthma; b) acute coronary artery disease; c) collagen vascular disease; d) pneumoconiosis; e) sarcoidosis; f) cancer; g) non-parenchymal restrictive lung disease; h) other severe comorbid conditions. In addition, patients who did not perform the home-based pulmonary rehabilitation program on a regular basis and those who voluntarily left the study were excluded.

The study protocol was approved by the local ethical committee and all patients gave their written informed consent.

Home-based pulmonary rehabilitation program

The patients were informed about the benefits and the importance of their adherence to the pulmonary rehabilitation program over the 12-week course. In order to increase patients' adherence to the program, a booklet was prepared and the instructions about the program were given to the patients with this booklet. Our home-based pulmonary rehabilitation program consisted of pursed-lips breathing, thoracic expansion exercises, diaphragmatic breathing exercises, upper and lower extremity exercises combined with breathing control (pectoral muscles stretch, trunk extension, bilateral shoulder elevation, sit-to-stand exercises using a chair) and a walking program (15-30 min/day). Breathing control training, coping strategies to deal with shortness of breath and relaxation training were given to the patients. The patients were instructed to perform all exercises five days a week, in three sessions per day with 10 repeats. They were recommended to have a break and rest in the case of excessive fatigue and shortness of breath and to continue the exercises according to their fatigue tolerance. The supervision of the program was done by phone calls once a week and a daily exercise query [6,8,14].

Outcome measures

All patients were evaluated with the same gain parameters before and after the 12-week long home-based pulmonary rehabilitation program. The perceived dyspnea severity during daily life activities was scored. Pulmonary function, exercise capacity, and quality of life of the patients were assessed.

Pulmonary Function Test (PFT): PFT was performed by an expert using a SensorMedics Vmax 22 machine (SensorMedics Inc., Anaheim, CA, USA) according to the ATS criteria [15]. Forced vital capacity (FVC), forced expiratory volume in 1 second (FEV_1), ratio of FEV_1 to FVC (FEV_1/FVC) and carbon monoxide diffusing capacity (DLCO) values were recorded [15].

Medical Research Council Scale (MRCs): this is a 5-point scale based on degrees of various physical activities that precipitate dyspnea. There are 5 stages of dyspnea severity in the scale. As the stage

progresses from 1 to 5, the severity of the perceived dyspnea increases. In our study, each patient was instructed to read the descriptive statements and then select the number which best matched his/her shortness of breath [16].

6-minute walking test (6MWD): The 6MWD was administered according to ATS criteria. At the end of the test, the distance walked in 6 minutes was recorded. Before and after the 6MWD heart rate, blood pressure, respiratory frequency, peripheral oxygen saturation, and severity of dyspnea were assessed [17]. The peripheral oxygen saturation (SpO₂) was recorded in sitting position using Palco 400 Model pulse oximeter (Palco Labs, Santa Cruz, USA). The severity of dyspnea was recorded using the Modified Borg Scale (MBS).

Modified Borg Scale (MBS): this is a 10-point scale with a non-linear scaling scheme using descriptive terms to anchor the responses from the participants (0 = no dyspnea, 10 = very severe dyspnea) [18].

Medical Outcomes Short-Form 36 (SF-36): General health-related quality of life was assessed with the SF-36. The SF-36 comprises eight multi-item dimensions, which are physical functioning, physical role (role limitations due to physical problems), vitality, social functioning, emotional role (role limitation due to emotional problems), bodily pain, general health, and mental health. Each of the dimensions is scored from 0 to 100, with higher scores indicating better health related quality of life [19]. The Turkish version of the SF-36 was used in our study [20].

Statistical analysis

Data were analyzed using SPSS (version 11.0). All values are shown as number, percentage or mean (standard deviation). Differences between assessment parameters before and after the home-based pulmonary rehabilitation program were compared using the Wilcoxon signed-rank test. A p value of 0.05 or less was considered statistically significant.

RESULTS

Seventeen IPF patients (mean age 62.8 ± 8.5 years) were enrolled according to the inclusion criteria during a one year period. A total of 15 patients completed the program. Two patients failed to complete the required number of sessions because of infectious disease and they were excluded from the study. The data analysis was performed on the 15 patients (5 female, 10 male) who completed the program. All patients stated that they had not experienced any problem such as dyspnea and fatigue when performing their exercises. The mean number of the weekly completed sessions was 13.2 ± 2.1 sessions.

Table I summarizes selected descriptive characteristics of the patients. Six of our patients (40%) had a positive smoking history. The mean cigarette consumption in these patients was 12.3 ± 7.4 pack/years. All patients were quitters during this study period. It was determined that the body mass index of the patients did not change during the 12-

week-long follow up (p = 0.51, Table I).

After the home-based exercise program, pulmonary function test results were not changed (p > 0.05). However, the perceived severity of dyspnea during daily life activities changed (p = 0.003, Table II).

A significant increase was found in 6MWD after the program (p = 0.04, Figure 1).

The perceived dyspnea, and leg fatigue severities were reduced, peripheral oxygen saturation was increased (p < 0.05, Figure 2a-b).

As the general health related quality of life of the patients was investigated, a significant improvement was observed in general health perception, physical role and emotional status subscores (p < 0.05, Table III).

DISCUSSION

Our study suggests that a home-based pulmonary rehabilitation program has positive effects on functional capacity in IPF patients. The results of our study show that the dyspnea and leg fatigue symptoms were reduced, while exercise capacity and general health related quality of life of the patients significantly increased after the 12-week-long home-based pulmonary rehabilitation program.

It has been reported in the literature that pulmonary rehabilitation increases exercise capacity and improves quality of life in restrictive pulmonary diseases; however, a standard program has not been yet recommended. In previous studies, it has been stated that the effects of different settings of rehabilitation programs, i.e. inpatient, outpatient, home-based or combined programs, on dyspnea and quality of life should be investigated [5,6,8,9,14]. Despite the positive effects of out-patient pulmonary rehabilitation programs in improving disease related functional losses in patients with COPD, access to hospital-based outpatient pulmonary rehabilitation is very limited world-wide and to maintain the positive effects of the pulmonary rehabilitation program is difficult. Thus

TABLE I: BASIC CHARACTERISTICS OF THE PATIENTS

Variable	Value*
Age, years	62.8 ± 8.5 (Range 49-75)
Sex, n = 15	5 female, 10 male
BMI, kg/m ²	Before the program 26.3 ± 3.8 p 0.51 After the program 26.5 ± 4.0
Disease duration, years	5.0 ± 3.8
Smoking history	40% (n = 6) positive
Cigarette consumption, pack/years	12.3 ± 7.4
Education level	≤ 8 years 73.3% (n = 11), 8 years or more 26.7% (n = 4)

*Values are presented as number, percentage or mean (SD).

Definition of the abbreviation: BMI: Body mass index.

TABLE II: PULMONARY FUNCTION TEST RESULTS AND MRCS SCORES

	Before the pulmonary rehabilitation program	After the pulmonary rehabilitation program	p
FEV ₁ , L	1.9 ± 0.5	2.1 ± 0.4	0.16
% pred	64.7 ± 7.2	66.5 ± 4.9	0.21
FVC, L	2.3 ± 0.8	2.4 ± 0.5	0.31
% pred	71.6 ± 8.2	73.9 ± 5.3	0.28
FEV ₁ /FVC, %	82.7 ± 5.6	85.6 ± 9.7)	0.09
DLCO, mL/min/mmHg	11.3 ± 4.9	11.2 ± 5.1	0.27
% pred	68.0 ± 32.3	68.2 ± 33.8	0.7
MRCS	2.3 ± 1.2	1.4 ± 1.3	0.003*

Values are presented as mean ± SD. * p ≤ 0.05.

Definition of abbreviations: DLCO, carbon monoxide diffusing capacity; FEV₁, forced expiratory volume in 1 second; FVC, forced vital capacity; MRCS, Medical Research Council Scale.

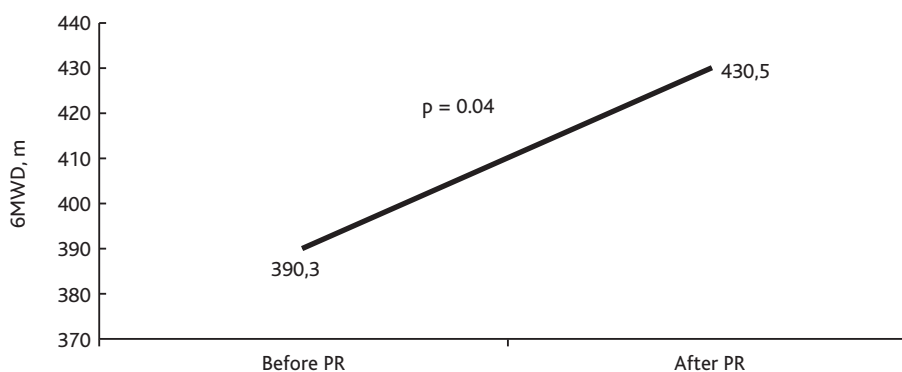
home-based rehabilitation programs have been accepted as an effective alternative to in/outpatient programs [11,12,21]. In Holland et al.'s study [9], an unsupervised home exercise program after the 8 weeks supervised exercise program was prescribed to idiopathic lung disease patients. However, the authors found that there was a decline in functional parameters. There is only one study addressing the effects of an 8-week home-based exercise program after the 4-week inpatient exercise program on dyspnea severity and quality of life in idiopathic lung disease patients [5]. Thus we believe that our study is of importance since it investigates the effects of a home-based pulmonary rehabilitation program on functional parameters in IPF patients.

Symptoms in patients with IPF may impair exercise capacity and quality of life. Therefore, an important aim of the assessment and treatment programs in IPF patients is symptom management. Dyspnea, fatigue, and cough have been shown to be the main IPF symptoms [1,2,22]. According to the above-mentioned aim, Nishiyama et al. [8] showed that their 10-week-long pulmonary rehabilitation program in patients with idiopathic pulmonary fibrosis

did not have an effect on respiratory functions and gas exchange but reduced dyspnea perception and improved exercise capacity. Holland et al. [9] reported that their 8-week-long supervised exercise training program performed in patients with idiopathic lung disease reduced disease symptoms and increased exercise capacity. Jastrzebski et al. [5] showed that a combined rehabilitation program (4 weeks inpatient and 8 weeks home-based) improved the quality of life and sensation of dyspnea in patients with idiopathic lung disease (21 with idiopathic pneumonia, 4 with lung fibrosis due to allergic alveolitis, 4 with lung fibrosis due to collagen disease, 2 with lung fibrosis due to silicosis), despite there being no changes in pulmonary function test results. Similarly, our study showed that the 12-week-long home-based pulmonary rehabilitation program performed in patients with IPF significantly reduced dyspnea severity during daily life activities, and dyspnea and leg fatigue severities after the 6MWD, but there was no improvement in respiratory function.

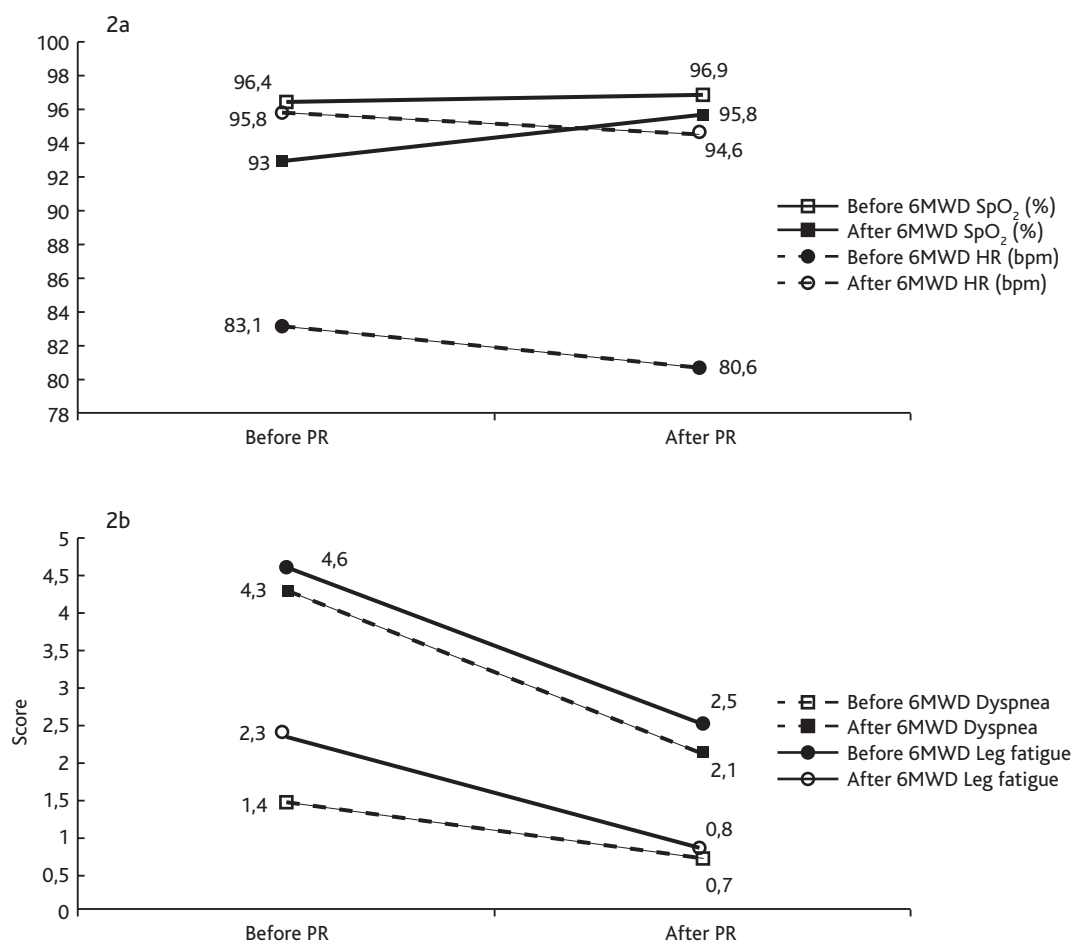
Ong et al. [14] reported that exercise capacity was increased significantly and effort dyspnea was

FIGURE 1: THE 6-MINUTE WALKING DISTANCE BEFORE AND AFTER THE HOME-BASED PULMONARY REHABILITATION PROGRAM



Definition of abbreviations: 6MWD, 6-minute walking distance; PR, pulmonary rehabilitation.

FIGURE 2: THE 6-MINUTE WALKING TEST RESULTS BEFORE AND AFTER THE HOME-BASED PULMONARY REHABILITATION PROGRAM



Definition of the abbreviations: 6MWD, 6-minute walking distance; bpm, beats per minute; HR, heart rate; PR, pulmonary rehabilitation; SpO₂, peripheral oxygen saturation

reduced after the 6-week-long outpatient pulmonary rehabilitation program in 34 patients with chronic lung diseases (COPD, idiopathic lung disease and bronchiectasis). Consistent with the above mentioned study, we determined a statistically significant increase in the exercise capacity of our patients after the home-based pulmonary rehabilitation program. The 6MWD increase was 45 meters in our study and it is similar to Naji et al.'s result [6]. This increase in our study is 10 meters greater than that in Holland et al.'s study, although our 6MWD increase is smaller than that reported in previous pulmonary rehabilitation studies performed in patients with COPD (< 54 meters) [9]. This difference could be related to the mixed content of other study groups. In our study, dyspnea and leg fatigue were reduced in a statistically significant manner after the home-based pulmonary rehabilitation program. According to these results, we considered that this increase in the 6MWD distance might be a result of the reduced disease symptoms, especially dyspnea and leg fatigue. In addition, the decrease in the fall of peripheral oxygen saturation after the home-based pulmonary rehabilitation program suggests that pulmonary functions improved after the

program. This might be an important functional gain parameter, because according to the literature, walking distance covered during the 6MWD, post-test change in desaturation and quadriceps muscle fatigue are associated with disease progression and mortality in IPF patients [8,23]. The aim of the pulmonary rehabilitation programs performed in individuals with idiopathic lung disease is to improve the health related quality of life as well as exercise capacity. It has been reported in the literature that exercise applications improve the quality of life in chronic respiratory disorders [2,5,14,24]. Jastrzebski et al. [5] showed that the 12-week long exercise program applied both on an inpatient and home basis improved dyspnea perception and quality of life assessed by SF-36 quality of life health survey. Similarly, general health related quality of life was assessed with the SF-36 quality of life health survey in our study and a significant increase was recorded in the scores for 'physical role', 'general health' and 'emotional role' categories. IPF is a chronic and progressive lung disease. The impaired physical fitness capacity may lead to reduced physical activity level in chronic lung dis-

TABLE III: THE GENERAL HEALTH RELATED QUALITY OF LIFE SCORES

Medical Outcomes Short-Form 36	Before the pulmonary rehabilitation program	After the pulmonary rehabilitation program	p
General health	57.0 ± 4.6	74.0 ± 4.7	0.04*
Physical function	56.0 ± 5.7	58.7 ± 7.3	0.24
Physical role	25.0 ± 1.7	68.3 ± 1.6	0.01*
Bodily pain	67.3 ± 2.6	72.0 ± 2.2	0.40
Vitality	52.0 ± 4.9	55.0 ± 4.2	0.40
Social function	75.8 ± 2.7	89.1 ± 1.8	0.17
Emotional role	29.0 ± 1.3	65.0 ± 1.4	0.02*
Mental health	49.9 ± 6.7	56.8 ± 5.4	0.14

Values are presented as mean ± SD.

* p < 0.05.

eases. Thus, maintaining the beneficial effects of the pulmonary rehabilitation programs is important [11,22]. Holland et al. [9] determined that the increase in exercise capacity acquired after their exercise program was not sustained for 6 months. Naji et al. [6] reported that the improvement in the exercise capacities of 46 patients with restrictive lung diseases (35 IPF patients and 11 patients with skeletal anomalies) was maintained for 1 year. However, in our study the long-term effects of the home-based pulmonary rehabilitation program was not investigated. In addition, the progression of the disease may lead to psychosocial disorders [25]. Thus, for future studies, the effects of home-based pulmonary rehabilitation programs on psychological problems and the long term effects of the home-based pulmonary rehabilitation programs should be assessed due to the progressive nature of IPF.

CONCLUSION

The results of this study show that a home-based pulmonary rehabilitation program may be beneficial to improve functional capacity of IPF patients by decreasing dyspnea perception, leg fatigue, increasing exercise capacity and improving general health related quality of life. Thus, we recommend that home-based pulmonary rehabilitation programs, which are tailored to the individual needs of IPF patients, should be included in the treatment plan. Nevertheless, further studies, featuring larger patient and control groups, are needed to confirm the effectiveness of home-based pulmonary rehabilitation in IPF.

CONFLICT OF INTEREST STATEMENT: None of the authors has any conflict of interest to declare in relation to the subject matter of this manuscript.

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