

# PRIMARY PROGRESSIVE FREEZING GAIT

## CASE REPORT

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**ABSTRACT** - The clinical features of the primary progressive freezing gait, a rare and distinct neurological entity, have been described in some articles. This paper describes this gait disorder affecting one patient, whose condition's long course provoked no postural instability. As stated in the literature, the pathophysiology of the primary progressive freezing gait still remains unclear, although the lack of response to levodopa therapy suggests a possible involvement of nondopaminergic pathways. Alteration in the visual perception may be a potential cause for the condition.

**KEY WORDS:** movement disorders, isolated freezing gait (primary, progressive).

### **Marcha congelada progressiva primária: relato de caso**

**RESUMO** - Uma rara e distinta entidade neurológica, a marcha congelada progressiva primária, tem sido relatada na literatura. Descrevemos o caso de um paciente com este distúrbio da marcha que, apesar do longo curso da doença, não apresentava instabilidade postural. Como citado na literatura, a fisiopatologia desta condição permanece obscura, embora a falta de resposta à levodopa sugira uma possível participação de vias não dopaminérgicas. Alteração na percepção visual também pode estar envolvida na gênese desta afecção.

**PALAVRAS CHAVE:** desordens do movimento, marcha congelada isolada (primária, progressiva).

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An isolated stubborn alteration of gait, with no signs of bradykinesia, rigidity, or tremor has been described in the literature as a "primary progressive freezing gait" (PPFG). This condition has been characterized as a hesitation to start walking, usually accompanied by festination, postural instability and recurrent falls that appear only while walking<sup>1</sup>. The condition also seems to be related to a distortion of unknown pathophysiology in relation to the visual perception of space<sup>9</sup>. A similar alteration was described by Atchison et al.<sup>2</sup> as "syndrome of gait ignition failure". However, this syndrome, differently from PPFG, presents normal equilibrium or slight alteration and no festination<sup>2</sup>.

This is a case study of a male patient, 73 years old, with a long-term history of gait alteration, without postural instability or any other signs of parkinsonism.

## CASE REPORT

A 73 year old man was evaluated during four years at the Movement Disorders Ambulatory, Department of Neurology, Hospital Universitário Pedro Ernesto, Universidade do Estado do Rio de Janeiro, Brazil. Nine years before, he noticed a slowly progressive change in his walk, characterized by some difficulty to lift the feet

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properly, which made him stumble whenever the floor surface was uneven. In addition, he found himself unable to go through doorways, arches, corridors or even walk among people and chairs that would narrow his passage. He reports that in such conditions he felt that the feet were glued to the floor. If there was any obstacle, he would freeze suddenly. None of these alterations happened when he walked in open places and/or backwards. A discrete improvement in gait could be observed when the patient walked along parallel lines marked on the floor, or after he drank small quantities of alcohol. He never complained of weakness in his lower limbs. He had no relevant past medical history nor a family history of any significance.

Parallel to a gait disorder, the patient reports a discrete slowness in some of his movements starting two years prior to the study. However, he did not notice any signs of worsening of this condition. During this time he reported the use of biperiden and later levodopa associated to carbidopa which did not improve his condition. He reports improvement with selegiline.

The neurological examination in the beginning of the study revealed the patient's difficulty to start walking whenever there were obstacles that diminished the physical space. In this situation he was not able to move the feet for approximately one minute. Sometimes he could lift the heels, but could not lift the tip of his toes. Each footstep measured 30 to 50 cm and there were no abnormalities in the associated movements of his upper limbs. After the initial difficulty, he was able to walk freely even when he changed his direction suddenly. He could also stand up from a chair without hesitation. His muscular strength was normal. A very slight rigidity was noticed at the right elbow and knee, but this condition did not worsen during the 4 years of follow-up study. No bradykinesia nor tremor were present; the postural reflexes were preserved and the deep reflexes were brisk in the four limbs. The plantar reflex appeared in flexion in both feet and palmomental reflex was elicited bilaterally. The mental examination showed no abnormalities.

The clinical evaluation and laboratory exams were normal. The MRI of the brain showed mild cortical atrophy, small hyperintense lesions in T2 in the periventricular and left cerebral white matter, and centrum semiovale. The electroencephalogram showed a slow alpha rhythm, 8 a 10 Hz, low voltage, with 6 a 7.5 Hz irregular waves, diffusely distributed and increased with hyperventilation.

## DISCUSSION

A person's life quality is undoubtedly influenced by his ability to move about firmly and independently. A person's gait involve an adequate musculoskeletal apparatus, supporting reflexes to maintain an upright posture, control of lateral and forward sway of the body, a motor program to perform repetitive, rhythmic movements which are originated in the spinal cord, in addition to the ability of adjusting the body to sudden or unexpected changes in direction. There is also an integrative system that selects the adequate patterns of muscle activation, according to the sensory input. This system is located in the brainstem, subcortical nuclei and frontal cortex, and generates the postural and motor responses. The motor cortex, basal ganglia and cerebellum can only modify the motor program<sup>10</sup>.

A typical gait is seen in the elderly and is called "senile gait pattern". It is characterized by a curved back, short steps, reduced velocity, increased time with both feet on the ground with no difficulty to start walking, and decreased hip rotation and associated upper limb swing. Elble et al.<sup>4</sup> believed that the short steps, due to a reduced muscular power of the hips and knees or to increased muscular stiffness, would explain the gait pattern of the elderly.

Fitzgerald and Jankovic<sup>7</sup> analyzed 10 patients with difficult walking and postural instability, with minimum or no involvement of the upper limbs. They called it "lower body parkinsonism." The authors observed initial hesitation, freezing, lack of festination and improvement with visual aid. They believe that the compromise of small vessels in their subjects led to a subcortical ischemia provoking the disconnection among the basal ganglia and the supplementary motor area which was then reported as the pathophysiological basis for the condition. All patients studied had multiple subcortical white matter lesions observed on MRI. This clinical condition differed from Parkinson's disease for its rapid course, lack of response to levodopa therapy and high prevalence of hypertension.

Many patients with gait disorders seek alternative means to overcome their difficulty. Some of the resorts used are stepping over the cane handle, walking along parallel lines on the floor and

other expedients that increase the complexity of the gait. It is supposed that these resorts that involve cortical mechanisms, not used in normal gait, modify the regular motor program which is defective in such patients and supply them with an alternative means to initiate and maintain motion<sup>10</sup>.

Mestre et al.<sup>9</sup> consider PPFG to be a sensory motor disturbance related to a visual alteration of the spatial perception associated or not to visual accuracy. The fact that, whenever blindfolded, the patients did not present freezing led them to conclude that freezing has a visual component.

According to Fitzgerald and Jankovic<sup>7</sup> and Fahn<sup>5</sup>, the lack of response to levodopa therapy suggests that freezing as an isolated phenomenon is not related to dopaminergic pathways. This is a plausible finding since neither the patients of Achiron et al.<sup>1</sup> nor ours showed an improvement with levodopa, even when low doses were administered.

Our patient did not show postural instability, frequent falls nor difficulty in changing directions abruptly. He did have festination and a great difficulty to start the gait and/or walk among people, and go through an arch. He was able to start walking when there were parallel lines drawn on the floor. He could surpass obstacles with his eyes closed and could easily walk backwards. These findings corroborate the hypothesis of the participation of visual spatial perception component in the pathophysiology of this condition<sup>9</sup>.

During the follow-up there were no signs of gait worsening nor other abnormalities of the neurological examination, except for senility signs (discrete elbow and knee rigidity), as suggested by other authors<sup>6</sup>. We were not able to establish a correlation between the neuroradiologic findings and the severity of the gait disability. According to Bradley et al.<sup>3</sup> and Hawke et al.<sup>8</sup>, patients over 50 years of age frequently present abnormal MRI's showing areas of hyperintensity in the white matter of hemispheres and periventricular regions without a morbid condition.

As it was mentioned, the variety of clinical differences of this condition and the fact that no specific anatomical alterations have been established have led us to conclude that more than one factor concur in the establishment of the pathophysiology of PPFG.

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