

Evaluation of Patients with Dysfunction of the Extra-Pyramidal System — Selection of Measuring Tools in Neurogeriatrics

Ocena stanu chorego z dysfunkcją układu pozapiramidowego — wybrane narzędzia pomiarowe w neurogeriatrii

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Summary

The group of diseases with extra-pyramidal disorders includes inter alia Parkinson's disease. It is one of the most common progressive, degenerative diseases of the central nervous system occurring in mature and older people. First symptoms of the disease occur in most cases after fifty years of age. The disease is generated by the loss of dopaminergic cells in the substantia nigra manifested clinically in the mobility slowdown, muscle rigidity and tremor at rest. The clinical use of various measuring tools (scales, indexes, indicators, tests, etc.) for the diagnosis of patients with dysfunction of the extra-pyramidal system has been increasingly applied. Based on the literature there were discussed scales most commonly used in the assessment of patients with dysfunction of the extra-pyramidal system including mainly those with Parkinson's disease. There were presented data regarding such measuring scales as: Hoehn and Yahr's (HY Scale), Webster's (Webster Rating Scale — WRS), Unified Parkinson's Disease Rating Scale (UPDRS), Northwestern University scale (Northwestern University Disability Scale — NUDS), Columbia University scale (Columbia University Rating Scale — CURS), Schwab and England's independence scale (Schwab and England ADL Scale — S-E ADL), the short scale of Parkinson's disease evaluation (Short Parkinson's Evaluation Scale — SPES), the scale of daily activities in Parkinson's disease (Parkinson's Disease Activities of Daily Living Scale — PDADL) as well as life quality scales, including the PDQL (Parkinson's Disease Quality of Life Questionnaire) and PDQ-39 (Parkinson's Disease Questionnaire). (*JNNN 2014;3(2):88–92*)

Key Words: assessment of patient's condition, extra-pyramidal system, scales, neurogeriatrics

Streszczenie

Do grupy schorzeń przebiegających z zaburzeniami układu pozapiramidowego należy między innymi choroba Parkinsona. Jest jedną z najczęstszych postępujących, zwyrodnieniowych chorób ośrodkowego układu nerwowego, występujących u ludzi dojrzałych i starszych. Pierwsze objawy schorzenia pojawiają się w większości przypadków po 50 roku życia. U podłoża schorzenia leży zanik komórek dopaminergicznych istoty czarnej przejawiający się klinicznie spowolnieniem ruchowym, sztywnością mięśni i drżeniem spoczynkowym. Wykorzystanie w praktyce klinicznej różnorodnych narzędzi pomiarowych (skal, indeksów, wskaźników, testów itp.) dla potrzeb diagnozowania pacjentów z dysfunkcją układu pozapiramidowego jest coraz częściej stosowane. W oparciu o piśmiennictwo omówione zostały najczęściej używane skale w ocenie pacjentów z dysfunkcją układu pozapiramidowego, w tym głównie z chorobą Parkinsona. Przedstawiono dane dotyczące takich skal pomiarowych, jak: skala Hoehna i Yahra (HY Scale), Webstera (Webster Rating Scale — WRS), Unified Parkinson's Disease Rating Scale (UPDRS), skala Uniwersytetu Northwestern (Northwestern University Disability Scale — NUDS), skala Uniwersytetu Columbia (Columbia University Rating Scale — CURS), skala samodzielności Schwaba i Anglii (Schwab and England ADL Scale — S-E ADL), krótka skala ewaluacji choroby Parkinsona (Short Parkinson's Evaluation Scale — SPES),

skala czynności życia codziennego w chorobie Parkinsona (Parkinsons Disease Activities of Daily Living Scale — PDADL) oraz skale oceny jakości życia, w tym skala PDQL (Parkinsons Disease Quality of Life Questionnaire) i PDQ-39 (Parkinsons Disease Questionnaire). (PNN 2014;3(2):88–92)

Słowa kluczowe: ocena stanu chorego, układ pozapiramidowy, skale, neurogeriatria

Introduction

The extra-pyramidal system is said to be one of main of central regulators of involuntary movements and muscular tone. By working with cerebellum, it modulates the process of involuntary movement conditioned on the extra-pyramidal system, guaranteeing its fluency, clarity, precision and optimal execution time. The group of diseases with the extra-pyramidal system disorders includes: Parkinson's disease, symptomatic Parkinson's syndromes, cerebral degenerative syndromes associated with Parkinson's disease, dementia and/or other symptoms associated ('parkinsonism plus') chorea, dystonia, tics disease and athetosis. Among the most common is Parkinson's disease.

Parkinson's disease is a progressive, degenerative disease of the extra-pyramidal system. It is one of the most common diseases of the central nervous system occurring in mature and older people. First syndromes of the disease occur in most cases after fifty years of age. The disease is generated by the loss of dopaminergic cells in the substantia nigra manifested clinically in the mobility slowdown, muscle rigidity and tremor at rest [1–5].

The most common initial symptom is a tremor of usually one hand, sometimes of both regarding fingers performing pills turning movements. In the beginning of the illness, tremor is usually single-sided. It occurs at the rest and may intensify under stress or due to tiredness as well as at low temperatures; it usually decreases during active, intended movements and it disappears during sleep. It is rhythmic, may cover upper or lower limbs, mouth, the tongue or the head. Parkinson tremor is observed in approximately 80% of patients already in the beginning of the development of stage of the disease; it often disappears at its later stage [6–8].

Parkinson rigidity is also typical for this disease and is manifested in the resistance at involuntary movements. Just as in the case of tremor, muscular tension is probably generated by the excess of acetylcholine. It has strongly generalized nature and can be most easily manifested by the movements related to binding and straightening the wrist or abduction and adduction movements. Most often it occurs more intensively in one half of the body. Increased muscle tension, is usually felt by patients usually as muscle stiffness in the arm or leg [6,8]. Sometimes increased tension of whole muscle groups is perceived as tiredness, fatigue or even pain. This symptom contributes to the change of patient's posture, creating characteristic picture of the posture of the patient

with Parkinson's disease: Upper and lower limbs slightly bent, arms extended forward, torso and head tilted forward and sometimes slightly swaying from side to side. Gait becomes shuffling, with short steps, and there is no corresponding movement of hands. Sometimes, unintentionally there may be occur acceleration of steps where the patients starts to run in order to avoid falling over. Posture disorders are visible in both the standing as well as sitting position. The head tends to fall forward on the chest and in the case of some patients the torso falls forward or backward if it is not supported [4,5,8]. The patients have a tendency to falling over which results from impaired reflexes of posture. In some patients there occur incidents of sudden interruption of the movement performed, so called solidification (*freezing*) consisting in the inability to detach feet from the ground while walking or movement initiating. It might result from stress and occurs spontaneously in narrow spaces and passageways in front of an obstacle and when entering an open space [6–9].

Another characteristic symptom which outlines the profile of a person with Parkinson's disease is the slowness of movement (bradykinesia). It refers to slowing voluntary movements, slow getting up from chairs, beds, getting dressed slowly or sluggish performance of other daily activities, the patient has problems with a simultaneous performance of two activities or with the performance of fast alternating movements. This problem also refers to the impoverishment of movement as compared to automatic movements performed unconsciously, this includes for example: blinking, movements of facial expression, swallowing saliva, balancing the upper limbs while walking. Slowing down and impoverishment of movements as well as increased muscle tension make the face and posture of the patient at the advanced stage of the disease form a typical 'Parkinson like' image. The face of the patient is poor in facial expressions and resembles a mask — the so called "Poker face", which due to the excessive seborrhea makes the impression of an oiled one [4–9].

Parkinson's disease is also characterized by the change of handwriting defined as micrograph [9]. It consists in the fact that written words and letters are becoming more and more illegible and are getting smaller, particularly in the end of the sentence. Sometimes one can see the evidence of hand shaking which makes handwriting still more illegible. Micrograph intensifies with the progress of the disease. The change is clearly visible when the form of patient's handwriting from a few years before —

from the period of health, is compared with the current handwriting. In some patients progress of the disease generates the occurrence of speech and swallowing disorders [7–9]. Speech gradually becomes more and more silent, monotonous, loses its melody and intonation and in some cases it gets so much accelerated that the patient is hardly understood by others [7,9]. Disorders in swallowing occur rather at later stages of the disease and result both from general bradykinesia as well as from autonomic disorders. The patient swallows more rarely and more slowly, the time for meal is considerably extended and as a result the patient avoids having solid food which becomes substituted by semi-liquid meals. Sometimes, swallowing disorders may lead to aspiration pneumonia or choking. Less frequent automatic movements of swallowing generate excess of saliva, which — particularly at night — leaks out of the mouth leaving traces on the pillow [8]. Quite often, in the case of Parkinson's disease there may also occur disorders in urination and defecation in the form of persistent constipation and pollakiuria, nycturia, urgency, and less frequently urinary incontinence [6,9]. As regards cardiovascular disorders, there most often occurs orthostatic hypotension which means decrease in blood pressure in the case of a sudden change in body position from lying or sitting to standing which may generate dizziness, fainting and collapsing. Hypotension most often takes place in the morning hours [6,8]. Taking into account the mental state, patients with Parkinson's disease very often suffer from depression [4,5,7,9]. Periods of sadness, depression or bad mood resulting from the lasting illness, which in some way interferes with and reduces patient's mobility, may be prolonged and might last for weeks and months. With the progression of disease and intensification of its symptoms the patient becomes increasingly dependent on his or her caregivers, whereas patient's deteriorated mobility in the final stage of the disease results in the loss of the ability to move and makes the patient stay permanently in bed or move only when accompanied by caregivers.

The most common scales and rates applied by therapeutic teams in the diagnosis of the condition of the patient with a dysfunction of the extra-pyramidal system, including Parkinson's disease, are: Hoehn i Yahr's scale (HY Scale), Webster's (Webster Rating Scale — WRS), Unified Parkinson's Disease Rating Scale (UPDRS), Northwestern University scale (Northwestern University Disability Scale — NUDS), Columbia University scale (Columbia University Rating Scale — CURS), Schwab and England's independence scale (Schwab and England ADL Scale — S-E ADL), the short scale of Parkinson's disease evaluation (Short Parkinson's Evaluation Scale — SPES), the scale of daily activities in Parkinson's disease (Parkinson's Disease Activities of Daily Living Scale — PDADL) as well as life quality

scales, including the PDQL (Parkinson's Disease Quality of Life Questionnaire) and PDQ-39 (Parkinson's Disease Questionnaire). (Parkinson's Disease Quality of Life Questionnaire) and PDQ-39 (Parkinson's Disease Questionnaire).

The most commonly applied measuring tools for the assessment of patients — review of literature

Hoehn and Yahr scale (HY Scale) (1967) [10] is one of the oldest and at the same time most commonly applied measuring tools for the assessment of the course and treatment of Parkinson's disease. The scale differentiates 5 degrees of the disease as stages of its progression, from degree I — single-sided damage, without functional disorders, to degree V — the sitting or lying patient, who requires assistance from third persons. This scale has been applied all over the world mainly in multicenter clinical tests. It is clear, widely applied and accepted.

Webster scale (Webster Rating Scale — WRS) (1968) [11] is a tool evaluating deficits as well as the degree of patient's disability. Ten parameters are evaluated in the four-point scale. 0 point means that there is no damage, 3 points mean that the damage is very serious. Based on the total score, the patient can be allocated to one of the groups of disability: group I (1–10 points) — early stage of the disease, group II (11–20 points) — moderate disability and group III (21–30 points) — advanced stage of the disease.

Unified Parkinson's Disease Rating Scale (Unified Parkinson's Disease Rating Scale (UPDRS) (1987) [12] is a specific tool used for the evaluation of the severity of the disease and for monitoring its course. In its classic version the scale consists of 6 integrally interconnected parts regarding: 1 — the intellectual status and mood disorders, 2 — daily activities, 3 — locomotor system test, 4 — analysis of treatment complications, 5 — evaluation of illness stage and 6 — assessment of independence. Due to numerous reports regarding its psychometric properties this scale has had many modifications.

Northwestern University Scale (Northwestern University Disability Scale — NUDS) (1961) [13] is used for the evaluation of patients suffering from Parkinson disease. It evaluates five parameters: 1 — walking, 2 — getting dressed, 3 — personal hygiene, 4 — having meals and 5 — speech. In the eleven-degree scale the patient receives 0 points, if he does not perform the given activity and 10 points if the patient's condition is correct.

Columbia University scale (Columbia University Rating Scale — CURS) (1993) [14] is considered to be the most accurate and homogeneous scale evaluating 14 symptoms of Parkinson disease. The patient may obtain

maximum 56 points when the five-point criterion (0–4) is applied.

The scale of daily activity of the patients suffering from Parkinson's disease (Schwab and England Activities of Daily Living S-E ADL) (1969) [15] is a simplified version of the scale prepared in 1956, in the period of first thalamotomy. It generally evaluates the degree of patient's independence from the environment. The evaluation is quoted in the percentage figures. The patient who is totally independent of the environment and performs all daily activities without any problems and slowdown receives 100%. The patient bedridden with autonomic dysfunction receives 0%. This scale is used as the criterion for assessing the severity and progression of Parkinson's disease. For the purpose of practical classification of the patient, the percentage scale, compliant with scale criterion proposed, is suggested.

The short scale of Parkinson's disease evaluation (Short Parkinson's Evaluation Scale — SPES) (2004) [16] is also considered as the scale of evaluating the effects of Parkinson's disease (Scales for Outcomes in Parkinson's Disease — SCOPA). In the four-point scoring there are 21 parameters assessed which are related mainly to mobility functions. The scale has 3 sub-scales regarding: 1 — evaluation of mobility, 2 — daily activities and 3 — motor complications.

The scale of daily activities in Parkinson's disease (Parkinson's Disease Activities of Daily Living Scale — PDADL) (2001) [17], is a specific scale for a subjective, general assessment of patient's independence regarding daily activities. In the 5-point scoring proposed, the following elements are evaluated: having meals, getting dressed, care, walking, changing places, bathing and the use of toilet.

Parkinson's disease Quality of Life Questionnaire (Parkinson's Disease Quality of Life Questionnaire — PDQL) (1996) [18] is one of the two (apart from Parkinson's Disease Questionnaire) most common tools evaluating the quality of life in Parkinson's disease. The scale covers 4 fields for evaluation: Parkinson symptoms, systemic symptoms, emotional realm and social functions. In total, the questionnaire contains 37 scoring items on a 5-point scale of response from constantly to never.

The Questionnaire in Parkinson's disease (Parkinson's Disease Questionnaire — PDQ-39) (1995) [19] evaluates 39 parameters grouped in 8 aspects regarding most specific issues of the quality of life. The scale is characterized by good psychometric properties, confirmed by multicentre studies, as well as by good utility in the evaluation of features specific for parkinsonism.

In patients with the dysfunction of extra-pyramidal system one should not forget of the evaluation of such areas as: typical motor functions, walking, the risk of falling down, involuntary movements, dyskinesia; cognitive function, depression, fatigue, sleep disturbances

for which there are used specific tools such as: modified UPDRS — MDS-UPDRS scale (Movement Disorder Society — sponsored revision of the Unified Parkinson's Disease Rating Scale), test Up&Go (Timed Up&Go Test), AIMS (Abnormal Involuntary Movement Scale), PDYS-26 (Parkinson's Disease Dyskinesia Scale), UDysRS (Unified Dyskinesia Rating Scale) as well as MMP (Mini Mental Parkinson), PFS (Parkinson Fatigue Scale), PDSS (PD Sleep Scale) and PSQI (Pittsburgh Sleep Quality Index).

None of the aforementioned scales is ideal, best and more effective in the diagnosis. There are inconsistent reports regarding the ease of applying the scales, scoring schemes and the moment of evaluation. Some studies emphasize the need to use multiple tools simultaneously — for the purpose of a complete diagnosis of the patient's condition.

According to the experts of this subject, regardless of their structural qualities, scales are not able to replace traditional neurological examination or comprehensive geriatric assessment, but they may be used as a complement in the assessment of neurological deficit in prognosis or monitoring the course of the disease and effects of treatment. Summing up the above considerations it must be concluded that among the aforementioned measuring tools the recommended ones include: Hoehn and Yahr scale (HY Scale), Unified Parkinsons Disease Rating Scale (UPDRS), Northwestern University scale (NUDS), Schwaba and England independence scale (S-E ADL), the short scale of Parkinson's disease evaluation (SPES) and Parkinson's Disease Questionnaire (PDQ-39).

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