Evaluation of the quality of life in patients with segmental dystonia

Procena kvaliteta života bolesnika sa segmentnom distonijom

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Abstract

Background/Aim. Segmental dystonia is an abnormal movement, characterized by involuntary, sustained and repetitive muscular contractions, causing twisting and abnormal posturing of two or more adjacent body parts. It is not a life-reducing condition, but it deteriorates physical, mental and social functioning. The aim of the study was to define the basic demographic and clinical characteristics of patients with segmental dystonia and to estimate their quality of life.

Methods. The study included patients treated at the Clinic for Neurology – Clinical Center of Serbia (Department for Involuntary Movements). The patients with idiopathic segmental dystonia fulfilled the following questionnaires: general questionnaire, standard questionnaire for estimation of the quality of life SF 36, a list of questionnaires related to disease, and social participation scales. Statistical analysis involving the methods of descriptive statistics and linear regression analysis was used for predictive values of the characteristics.

Results. The study included 28 patients with segmental dystonia, the mean age of 53.1 ± 15.8 years. Analysis of SF 36 questionnaire item domains showed that patients with segmental dystonia had the lowest score in the domain of body pain (30.6 ± 28.2) and the highest in the domain of physical function (73.6 ± 19.6). Higher values of the scale of the disease severity (β = -0.526, 95% CI -4.719, -0.996; p = 0.0004) and Hamilton depression scale (β = -0.498, 95% CI -1.295, -0.227; p = 0.0007) were more significant predictors of low quality of life. Higher value of the Leisure activities scale (β = 0.611, 95% CI 0.242, 0.772; p = 0.001) was a significant predictor of better quality of life.

Conclusion. The most important predictors of low quality of life in patients with segmental dystonia were disease severity, low acceptance of illness, depression and low self-esteem.

Key words: dystonia; quality of life; questionnaires; sensitivity and specificity.

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DOI: 10.2298/VSP1209759B
Introduction

Dystonia is characterized by sustained and involuntary muscular contractions, causing twisting, repetitive movements and abnormal posturing of a body. According to distribution of involuntary movements, dystonia is divided to focal, segmental, multifocal, hemi- and generalized. Focal dystonia assumes that a single muscle or a muscular group is involved. Segmental dystonia involves two or more adjacent body parts and may be: cranial – two or more parts of the cranial or cervical musculature are involved; axial – trunk and neck are involved; brachial – either one arm and axial musculature or both arms with or without the neck and trunk are being involved, and crural – either one leg and trunk or both legs with or without the truncal involvement. The onset of segmental dystonia is most often focal, that is, a group of muscles is initially involved, with the progression of disease being observed in 15%-35% of patients, usually affecting the adjacent body part. The prevalence of different forms of late-onset focal dystonia ranges from 2 to 57 patients per million inhabitants, and it is estimated that the proportion of segmental dystonia accounts for 2%-20% of a total number of late-onset dystonias.

Dystonia is not life-reducing, nevertheless the affected people having these involuntary movements are faced with the multiple problems related to physical, mental and social health. It is a disease appearing in younger or middle-aged people, i.e. in their prime of familiarity, professional and life expectations in general. The disease is “visible”, cannot be self-controlled, and more or less, stigmatizes the affected people. In a significant number of patients, it is followed by pain and tremor, depending upon the extent of localization of the involuntary movements, it results in functional incapacity of varying degree. Ben-Sholomo et al. analyzing the quality of life in 289 patients with torticollis, found a surprising discrepancy between preserved physical status of patients on one hand, and their poor mental health and emotional state on the other. These results suggest that the quality of life in dystonic patients is not exclusively determined by the presence of involuntary movements, but also by other clinical, social, demographic and psychological factors, as well as their interaction.

The assessment value of the quality of life is based on the fact that therapeutical procedures are not simply intended for elimination or abatement of symptoms, but they also imply the assistance rendered to patients for as much as optimal living with their disease. On the other hand, measurement of the quality of life together with other clinical indicators allows for evaluation of the effectiveness of the applied therapeutical procedures.

The objective of our study was to define basic demographic and clinical characteristics of patients with segmental dystonia, as well as to evaluate the quality of life of these patients considering the effect of different demographic, clinical, psychological, social and other factors.

Methods

The study group consisted of patients treated at the Clinic of Neurology, Clinical Center of Serbia (Department for Involuntary Movements), who were diagnosed with dystonia in the period from 1990 to 2005. Inclusion criteria were as follows: present segmental dystonia with all clinical characteristics of idiopathic dystonias, examined and confirmed by two independent neurologists; normal findings of radiological visualization methods (computerized tomography – CT and or nuclear magnetic resonance – NMR) aiming at ruling out the structural lesion of the central nervous system as the cause of dystonia; normal results of laboratory tests (serum copper and ceruloplasmin, urine copper, acanthocytes in peripheral blood smear, immunoserological tests) with a view to exclude most frequent metabolic, degenerative or immunological causes of dystonia.

The patients who met the required criteria fulfilled the following questionnaires: General Questionnaire, Standard Questionnaire for Estimation of the Quality of Life SF 36, a list of questionnaires related to disease and Social participation scales.

The General Questionnaire was created for collection of basic demographic and clinical data, such as sex, age, qualification, marital status, age at the onset of disease, way and time of disease progression, presence of pain, tremor or other involuntary movements, modes of treatment and their effectiveness, etc. This questionnaire was designed to evaluate the severity of the disease by a patient’s self-estimation on the 1–10 scale (higher score—more severe disease).

The Generic Questionnaire for Estimation of the Quality of Life or Short-Form Health Survey (SF-26) is a scale proved to be very sensitive for evaluation of the quality of life, that is, severity of global effect of disease on the affected individual. This questionnaire covers eight health domains, divided into the following subscales: physical activities, physical activities and performance of everyday activities, general health, vitality, social activities, emotional activities and mental health. This study used the questionnaire version which was validated and adapted to Serbian-speaking region. SF-36 scoring of the results was carried out by the Likert method, 0 designating the worse and 100 the best possible health.

The disease-specific questionnaires include different scales evaluating the effect of internal factors to the quality of life, i.e. variables reflecting the premorbid characteristics of individuals, the way the disease is accepted, defense mechanisms, pattern of behavior as a response to the disease as well as the possibility of reactive depression and anxiety. For testing the abovementioned functions, the following scales were used: Rosenberg self-esteem and self-accusation scale, Felton acceptance of illness scale, Stigma scale, Hamilton depression scale, and Hamilton anxiety scale.

The social participation scales estimate the effect of cohabitation and different aspects of social support rendered to a patient by his/her cohabitants on the quality of life. To evaluate this aspect of the quality of life, the following scales were applied: Zimet multidimensional scale of perceived social support and Leisure time scale.

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Statistical analyses included descriptive methods (arithmetic mean, standard deviation and standard error). Linear regression analysis was used for analyzing the predictive values of variables.

Results

The our study included 28 patients with segmental dystonia whose basic demographic characteristics are presented in Table 1.

The eyelids were first affected by dystonic involuntary movements in 10/28 (35.7%) of the patients, the neck in 12/28 (42.9%), the right hand in 5/28 (17.9%) and the left hand in 1/28 (3.5%). After the involvement of the first region, the disease spreaded to another adjacent body parts in 26/28 (92.9%) of the patients during the mean time of 3.28 years (ranging from 10 days to 15 years). In 2/28 (7.1%) of the patients, the disease involved two regions from the very beginning, in one patient the right and the left hand simultaneously, and in another the eyelids and the lower face.

Table 2 illustrates the basic clinical characteristics of the patients with segmental dystonia. Tremor at the site of dystonia was evident in 13/28 (46.4%) and pain at the site of dystonic movement/position was present in 17/28 (60.7%) of the patients. Manifestation of other chronic diseases was reported in 12 (42.9%) of the patients with segmental dystonias. Botulin toxin injections were used for treatment in 15 (53.6%) of the cases, the average number of applications was 3.0 while the mean percent of improvement was 54.0%.

Table 3 illustrates the mean values of specific SF-36 questionnaire domains in patients with segmental dystonia. The patients with segmental dystonia had the lowest score in domain of body pain (30.6 ± 28.2) and the highest in the domain of physical function (73.6 ± 19.6).

Table 4 presents the mean values of different scales used for analysis of the quality of life in the patients with segmental dystonia.

The significance of demographic factors, such as sex, age at the onset of the disease, current age, education, employment and marital status for the quality of life was presented in Table 5, while the impact of clinical factors (duration of disease, tremor, pain, comorbidity and botulin toxin treatment) was illustrated in Table 6. None of these factors had significant effect on the quality of life in this group of the patients.

Table 7 presents the mean values of different scales used for evaluation of the quality of life in the patients with segmental dystonia was given in Table 7. Higher values of the Scale of disease severity (β = -0.526, 95% CI -4.719, -0.996; p = 0.0004) and Hamilton depression scale (β = -0.498, 95% CI -1.295, -0.227; p = 0.0007) were more significant predictors of the low quality of life. Higher value of the Leisure activities
scale ($\beta = 0.611, 95\% \text{ CI} 0.772; p = 0.001$) was a significant predictor of better quality of life in the patients with segmental dystonia. The values of Acceptance of Illness scale ($p = 0.0060$) and Self-Esteem scale ($p = 0.053$) were close to statistical significance.

**Discussion**

A total of 28 patients with segmental dystonia were analyzed, having focal disease in over 90\% of the cases, which subsequently spread to adjacent regions, giving the image of segmental form. In only 7\% of the examined patients, the nature of disease was segmental. At the onset of the disease, our patients were 43.3 (4–75) years old. Although dystonia may appear in any age, in 79\%–90\% of the time it is manifested between the fourth and sixth decade\(^9, 20–23\). The course of dystonia is unpredictable. The spread of dystonia is, before all, determined by age and location of the initial disorders; therefore, younger people and the onset of dystonia in legs increase not only the possibility of spreading but also the rate of expansion. Adult-age dystonia tends to progress lesser, and it usually develops several years from the initial manifestations\(^5\). The highest risk of the disease spreading to other body parts appears during the first

**Table 4**

| Scales                       | Values ($\bar{x} \pm \text{SD}$) |
|------------------------------|----------------------------------|
| Hamilton depression scale    | 7.2 ± 6.1                        |
| Hamilton anxiety scale       | 8.7 ± 5.8                        |
| Scale of disease severity    | 5.4 ± 1.7                        |
| Leisure activities scale     | 28.0 ± 1.2                       |
| Scale of acceptance of illness | 2.9 ± 0.7                      |
| Stigma scale                 | 14.4 ± 2.7                       |
| Multidimensional scale of personal support | 72.3 ± 10.0 |
| Self-esteem scale            | 15.0 ± 2.3                       |
| Self-accusation scale        | 12.0 ± 2.8                       |

**Table 5**

| Demographic factors | $\beta$ coefficient | 95\% confidence interval | $p$  |
|---------------------|---------------------|--------------------------|------|
| Sex                 | -0.085              | -9.189–5.992             | 0.669|
| Age at the onset of disease | -0.262         | -0.355–0.069             | 0.178|
| Actual age          | -0.131              | -0.355–0.069             | 0.506|
| Education           | 0.200               | -2.644–8.071             | 0.307|
| Employment          | -0.078              | -8.884–5.991             | 0.693|
| Marital status      | 0.231               | -3.890–15.014            | 0.237|

**Table 6**

| Clinical factors                  | $\beta$ coefficient | 95\% confidence interval | $p$  |
|-----------------------------------|---------------------|--------------------------|------|
| Duration of disease               | 0.311               | -0.086–0.828             | 0.107|
| Tremor at the site of dystonia    | -0.212              | -11.208–3.374            | 0.280|
| Pain at the site of dystonia      | -0.019              | -7.980–7.253             | 0.923|
| Other chronic diseases            | -0.044              | -8.343–6.688             | 0.823|
| Botulin toxin treatment           | 0.235               | -2.893–11.608            | 0.228|

**Table 7**

| Scales                                    | $\beta$ coefficient | 95\% confidence interval | $p$  |
|-------------------------------------------|---------------------|--------------------------|------|
| Hamilton depression scale                 | -0.498              | -1.295–0.227             | 0.007*|
| Hamilton anxiety scale                    | -0.307              | -1.118–0.124             | 0.112 |
| Scale of disease severity                | -0.525              | -4.719–0.996             | 0.004*|
| Leisure activities scale                 | 0.611               | 0.242–0.772              | 0.001*|
| Scale of acceptance of illness           | -0.359              | -7.392–0.171             | 0.060 |
| Stigma scale                             | -0.105              | -1.693–0.991             | 0.328 |
| Multidimensional scale of personal support| 0.192               | -0.192–0.555             | 0.328 |
| Self-esteem scale                        | 0.369               | -0.023–2.889             | 0.053 |
| Self-accusation scale                    | -0.148              | -1.744–0.802             | 0.454 |

* – statistically significant
five years. The study performed by Svetel et al. reported that the highest risk of dystonia spreading was in patients with the initial blepharospasm followed by patients with hand dystonia, torticollis and spasmodic dysphonia. The same study showed that the blepharospasm was also associated with the highest rate of disease progression. Defazio et al. in their study including 159 patients with the blepharospasm as the initial manifestation, found that the risk of spreading was independently increased by older age at the onset of disease, female sex and head and face injuries followed by loss of consciousness.

The quality of life is a complex category determined by individual and/or mutual action of different factors, such as demographic, social and premorbid, as well as factors related to specific characteristics of the disease itself. There are certain differences in determinants of the quality of life between some types of primary focal dystonias, but the question is how much distribution of these difficulties interferes with the quality of life of patients. Zetterberg et al. in their study in 2008 reported that the quality of life was most strongly associated with the level of physical activity and satisfaction as well as with the efficiency of the applied therapy, but that there was no significant correlation of the quality of life and the form of dystonia. The studies comparing the results of SF-36 questionnaire in dystonic patients with the general population criteria revealed that dystonic patients had significantly poorer quality of life in all domains, especially in those associated with physical and social functioning. In our study, none of demographic or clinical factors had significant influence on the quality of life in the patients with segmental dystonia, but depression significantly reduced the quality of life. The research conducted in our country to analyze the quality of life in 153 patients with different forms of focal idiopathic dystonias, showed that pain at the site of dystonic movement/position, depression and anxiety most significantly reduced the quality of life in patients with segmental dystonia, but depression significantly reduced the quality of life in the patients. The study was supported by the Ministry of Science and Technological Development of the Republic of Serbia (Grants no. 175090 and 175087).

The values of the Scale of acceptance of illness and Self-esteem scale were close to statistical significance, and a large-sample study would certainly show higher significance. Regardless of the application of different tests and methodology, the majority of studies suggest that the quality of life in patients with different forms of dystonia is determined, before all, by the presence of depression and anxiety, severity of disease and its stigmatizing effect. Dystonia spreading to different body parts does not necessarily mean a more severe form of disease, but it certainly leads to higher stigmatization and more difficult treatment of patients, thus to negative influence on the quality of life.

Conclusion

Regardless of the application of different tests and methodology, the majority of studies suggest that the quality of life in patients with different forms of dystonia is determined, before all, by the presence of depression and anxiety, severity of disease and its stigmatizing effect. Dystonia spreading to different body parts does not necessarily mean a more severe form of disease, but it certainly leads to higher stigmatization and more difficult treatment of patients, thus to negative influence on the quality of life.

Acknowledgment

The study was supported by the Ministry of Science and Technological Development of the Republic of Serbia (Grants no. 175090 and 175087).
REFERENCES

1. Fahn S. Concept and classification of dystonia. Adv Neurol 1988; 50: 1–8.
2. Jankovic J, Tolosa E. Dystonic disorders. In: Jankovic J, Tolosa E, editors. Parkinson’s disease and movement disorders. Baltimore: Williams & Wilkins 1998. p. 513–51.
3. Jahanshahi M, Martin MH, Marsden CD. Natural history of adult-onset idiopathic torticollis. Arch Neurol 1990; 47(5): 546–52.
4. Greene P, Kang U, Fahn S. Spread of symptoms in idiopathic torsion dystonia. Mov Disord 1995; 10(2): 143–52.
5. Tolosa E, Marti J. Adult-onset idiopathic dystonias. In: Watts R, Koller W, editors. Movement disorders-neurological principles and practice. New York: McGraw Hill 1996. p. 428–41.
6. De Fazio G, Berardelli A, Abbruzzese G, Coviello V, Cardella F, De Berardinis MT, et al. Risk factors for spread of primary adult onset blepharospasm: a multicentre investigation of the Italian movement disorders study group. J Neurol Neurosurg Psychiatry 1999; 67(5): 613–9.
7. Castelon Konkiewitz E, Trender-Gerhard I, Kamm C, Warner T, Ben-Shlomo Y, Camfield L, Warner T; ESDE collaborative group. Prevalence of primary late-onset focal dystonia in the Belgrade population. Mov Disord 2003; 18(11): 1389–92.
8. Matsumoto S, Nishimura M, Shibasaki H, Kaji R. Epidemiology of primary dystonias in Japan: comparison with Western countries. Mov Disord 2003; 18(10): 1196–7.
9. Ben-Shlomo Y, Camfield L, Warner T; ESDE collaborative group. What are the determinants of quality of life in people with cervical dystonia? J Neurol Neurosurg Psychiatry 2002; 72(5): 36–41.
10. Ware JE Jr, Snow KK, Gandek B. SF-36 health survey. Manual and interpretation guide. Boston, MA: The Health Institute, New England Medical Center; 1993.
11. Rosenberg M. Society and the adolescent self-image. Princeton, NJ: Princeton University Press; 1965.
12. Featon BJ, Revenson TA. Coping with chronic illness: a study of illness controllability and the influence of coping strategies on psychological adjustment. J Consult Clin Psychol 1984; 52(3): 343–53.
13. MacDonald LD, Anderson HR. Stigma in patients with rectal cancer: a community study. J Epidemiol Community Health 1984; 38(4): 284–90.
14. Hamilton M. A rating scale for depression. J Neurol Neurosurg Psychiatry 1960; 23: 56–62.
15. Hamilton M. Diagnosis and rating of anxiety. Br J Psychiatry Spec Publ 1969; 3: 76–9.
16. Zinnet CD, Powell SS, Farley GK, Workman S, Berkoff K.A. Psychometric characteristics of the Multidimensional Scale of Perceived Social Support. J Pers Assess 1990; 55(3–4): 610–7.
17. Kelly JR, Steinkamp MW, Kelly JR. Later-life satisfaction: does leisure contribute? Leisure Sci 1987; 9: 189–200.
18. Ronohot P, Marshall MP, Deliatolas G. Spasmodic torticollis review of 220 patients. Can J Neurol Sci 1991; 18(2): 143–51.
19. Duane DD. Spasmodic torticollis: clinical and biological features and their implications for focal dystonia. Adv Neurol 1988; 50: 473–92.
20. Chan J, Brin MF, Fahn S. Idiopathic cervical dystonia: clinical characteristics. Mov Disord 1991; 6(2): 119–26.
21. Jankovic J, Leder S, Warner D, Schwartz K. Cervical dystonia: clinical findings and associated movement disorders. Neurology 1991; 41(7): 1088–91.
22. Abbruzzese G, Berardelli A, Girlanfa P, Marsden R, Martinus D, Morante F, et al. Long-term assessment of the risk of spread in primary late-onset focal dystonia. J Neurol Neurosurg Psychiatry 2008; 79(4): 392–6.
23. Svetel M, Pekmezovic T, Jovic J, Ivonovic N, Draganovic N, Marij J, et al. Spread of primary dystonia in relation to initially affected region. J Neurol 2007; 254(7): 879–83.
24. Zetterberg L, Aqulonius SM, Lindmark B. Impact of dystonia on quality of life and health in a Swedish population. Acta Neurol Scand 2009; 119(6): 376–82.
25. Camfield L, Ben-Shlomo Y, Warner TT. Impact of cervical dystonia on quality of life. Mov Disord 2002; 17(4): 838–41.
26. Page D, Butler A, Jahanshahi M. Quality of life in focal, segmental, and generalized dystonia. Mov Disord 2007; 22(3): 341–7.
27. Lim VK. Health related quality of life in patients with dystonia and their caregivers in New Zealand and Australia. Mov Disord 2007; 22(7): 998–1003.
28. Ivonovic N. The determinants of quality of life in patients with idiopathic dystonia [dissertation]. Belgrade: School of Medicine; 2009 [Serbian].
29. Pekmezovic T, Svetel M, Ivonovic N, Draganovic N, Petronic I, Trpavovic DK, et al. Quality of life in patients with focal dystonia. Clin Neurol Neurosurg 2009; 111(2): 161–4.
30. Gudex CM, Hawthorne MR, Butler AG, Duffey P. Effect of dystonia and botulinum toxin treatment on health-related quality of life. Mov Disord 1998; 13(6): 941–6.
31. Lacey JH, Bithell SA. Body image and its disturbances. J Psychosom Res 1986; 30(6): 623–31.
32. Harris DL. The symptomatology of abnormal appearance: an anecdotal survey. Br J Plast Surg 1982; 35(3): 312–23.
33. Papathanasiou I, MacDonald L, Whurr R, Jahanshahi M. Perceived stigma in Spasmodic Torticollis. Mov Disord 2001; 16(2): 280–5.

Received on September 24, 2010. Accepted on March 8, 2011.

Basurović M, et al. Vojnosanitetski Pregl 2012; 69(9): 759–764.