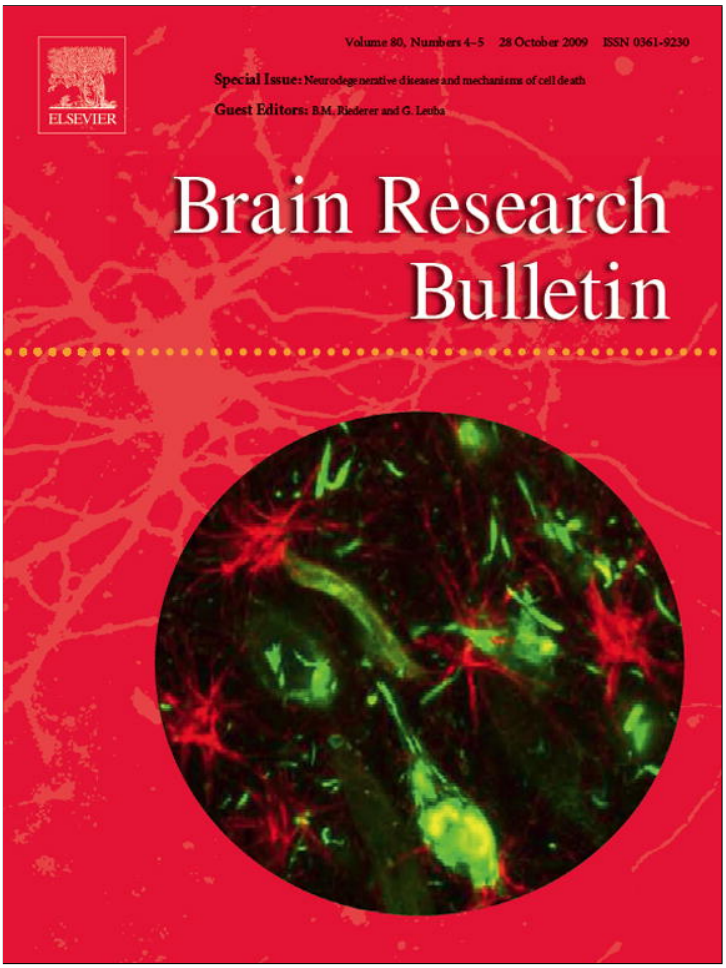
Provided for non-commercial research and education use.
Not for reproduction, distribution or commercial use.



This article appeared in a journal published by Elsevier. The attached copy is furnished to the author for internal non-commercial research and education use, including for instruction at the authors institution and sharing with colleagues.

Other uses, including reproduction and distribution, or selling or licensing copies, or posting to personal, institutional or third party websites are prohibited.

In most cases authors are permitted to post their version of the article (e.g. in Word or Tex form) to their personal website or institutional repository. Authors requiring further information regarding Elsevier's archiving and manuscript policies are encouraged to visit:

http://www.elsevier.com/copyright

Author's personal copy

Brain Research Bulletin 80 (2009) 192–195

Contents lists available at ScienceDirect

Brain Research Bulletin

journal homepage: www.elsevier.com/locate/brainresbull

Commentary

Coordinated multidisciplinary care for Huntington's disease. An outpatient department

Ruth B. Veenhuizen[a,*], Aad Tibben[b]

[a] Noorderbreedte persoonlijke zorg en wooncomfort, De Waring 6, 9001HZ Grou, The Netherlands
[b] Centre for Human and Clinical Genetics, Leiden University Medical Centre, P.O. Box 9600, 2300RC Leiden, The Netherlands

ARTICLE INFO

Article history:
Received 8 April 2009
Received in revised form 22 June 2009
Accepted 24 June 2009
Available online 1 July 2009

Keywords:
Huntington's disease
Multidisciplinary care
Coordinated care
Multidisciplinary team
Outreaching care

ABSTRACT

Huntington's disease is characterised by a complex set of motor, neuropsychologic and psychiatric symptoms which start slowly and progress over many years to a state of complete dependency. The symptomatic treatment during the ambulatory years is divers. In the northern part of the Netherlands coordinated multidisciplinary care is offered to patients diagnosed with Huntington's disease. A team of a neurologist, psychologist, occupational therapist, speech and language therapist, social worker and nursing home doctor monitors the patient and companion on a half-year basis and provide them with a plan of care. A case manager coordinates the plan of care in the dwelling place of the patient. In this article this type of care is outlined and illustrated with case histories.

© 2009 Elsevier Inc. All rights reserved.

1. Introduction

Huntington's disease (HD) is a dominantly inherited neurodegenerative disorder with a slowly progressive midlife onset. The HD mutation is a CAG repeat expansion [17]. The symptoms of HD comprise: involuntary choreic movements, decelerated intentional movements, personality changes, psychiatric symptoms and cognitive decline [11]. To this day, no cure has been found. From onset to complete care dependency takes about 15–30 years [6,7]. In the Netherlands, the majority of patients spend their final years in a nursing home. Although care programmes are available for the final stages [7,12], there is a lack of structured care pathways during the preceding years, in which patients live at home with the support of their families.

2. The need for a specialised, multidisciplinary approach

The nature of Huntington's disease, with its complex set of symptoms, requires specialised professional management [13]. Psychological and psychiatric symptoms range from emotional flattening, apathy, rigidity and irritability to depression, aggression, psychosis and obsessive-compulsive disorders [19]. Motor symptoms comprise choreic movements and delayed intentional movements, of which the latter are related to functional capacity [20]. Swallowing problems are among the high-risk consequences. Speech problems arise due to motor symptoms, impairment of processing information and retrieving stored knowledge. Other features of diminishing cognition are problems in visuospatial orientation, recognition of facial expressions, planning and initiation of activities [1]. Speech problems and unexpected social behaviour may lead to underestimation of neuropsychological capacities. On the other hand, anosognosia (lack of awareness of disease symptoms), prolonged intact memory and general intellect may lead to overestimation of the patient's capacities [5]. Besides these neurocognitive symptoms, most patients experience somatic symptoms during the course of the disease, like loss of weight, sweating, gastrointestinal disorders or micturition problems.

During the phase in which these symptoms become manifest, a multidisciplinary team can be built around the patient and may include the following professionals: neuropsychologist, neurologist, psychiatrist, psychiatric nurse, physiotherapist, speech and language therapist, occupational therapist, social worker, internist, dietician etc. As the family is just as well affected by the disease in daily routine, communication and social network, the family doctor, the lay organization and welfare workers should also play a role in keeping the patient's system in acceptable shape. Multidisciplinary care is a logical and compelling approach for a complex disorder like HD and several authors have emphasised the need for such care. For this type of care, various terms are used: multidisciplinary approach [1,8,14,4], comprehensive care [9], palliative care [10] and supportive care [2,15]. The underlying agreement on

* Corresponding author. Tel.: +31 621837331.
E-mail addresses: r.veenhuizen@chello.nl (R.B. Veenhuizen), a.tibben@lumc.nl (A. Tibben).

0361-9230/$ – see front matter © 2009 Elsevier Inc. All rights reserved.
doi:10.1016/j.brainresbull.2009.06.017

Author's personal copy

the contents and characteristics of HD patients care include proactive and long-lasting management, aiming at sustained functional capacities because of the progressive degenerative nature of the disease. All authors emphasise a practical approach, a flexible attitude, aiming for quality of life, with a lot of attention for family and psychosocial consequences. Considering the complexity of HD, care must be specialised and prepared for crisis management. Most authors stress the importance of education and research as part of care for HD patients. In this article we present a care system that seeks to meet the needs of patients diagnosed with Huntington's disease and for whom residential care is not yet necessary.

3. A multidisciplinary approach

3.1. The background

The background to the creation of an outpatient department for the assessment and treatment of HD patients in the northern part of the Netherlands was formed by the following case.

A 37-year-old man with symptomatic HD suffered from an obsession with defaecation, which resulted in a rectal prolapse and faecal spill all over the place. Because of this problem a colostomy was needed. The patient refused to go to a nursing home for rehabilitation. He lived on his own and his mother and sister had to work endlessly to keep his house clean. He drank too much and spent more than he could afford. He used to jump off his bicycle whenever he needed to stop instead of using the brakes, which caused dangerous situations. His social circle was small and his relatives were exhausted from his resistant attitude. The district nursing team experienced problems when helping him take care of his stoma and asked the nursing home doctor for support. Together with a case manager, a care plan was made for all the above-mentioned problems. A speech therapist, social worker, psychiatric nurse and occupational therapist were added to the team. Within a few months the plan started to work out well. The patient gradually accepted the day care treatment. The social worker took care of his financial problems and she even supported the entire family. The occupational therapist taught the patient how to ride a tricycle. A psychiatric nurse helped the patient to control his drinking habits and the nurses were trained how to deal with his defensive attitude. All these professionals worked according to one care plan and coordinated their interventions in multidisciplinary meetings. The nursing home doctor was responsible for the contents of the care plan and the case manager for the organization.

The effectiveness achieved through this collaboration, originated in the idea that quality of life of HD patients and their families could be improved in this stage of the disease and admission to a residential setting could be postponed potentially.

3.2. Establishing an outpatient clinic

In the Netherlands, a few categorical nursing homes for specialised HD care are set up, including day care facilities. Besides these residential settings, there are two specialised academic outpatient clinics for ambulatory HD patients. These clinics are at large distance from the northern part of the country. The day-to-day care for an ambulatory patient cannot easily be coordinated from these academic hospitals. This local lack of adequate care for HD patients together with the experience of the above-mentioned patient resulted in a project plan for an outpatient department for HD patients as a division of a health care institution. Funding of the project was found within the Dutch health insurance companies through innovation of care ordered by the Dutch Health Care Authority. The local health insurance company together with the umbrella organization of the local nursing home and the province agreed on funding the extra costs of the project for 2 years. Within this project 20 new patients can be seen each year. The specialised HD outpatient department is arranged like a carousel (see Fig. 1). The patient and companion are invited for an assessment that takes half a day. After a short introduction by the nursing home doctor, they visit the neurologist together. Subsequently, the patient and caregiver each visit the following professionals separately in different sequence: speech and language therapist, occupational therapist, nursing home physician, psychologist and social worker. At noon, the speech therapist and the occupational therapist will have lunch together with the patient and caregiver for observations on praxis and swallowing in a more relaxed environment. Thereafter, the multidisciplinary team discusses a care plan, which is presented to the patient and companions by the nursing home physician and the case manager. After mutual consent, a home implementation plan is figured out. The suggested care plan can involve home treatment, day care treatment, referral to a specialist, education or advice to district nurses or general physician etc. The care plan is reassessed every 6 months with a similar assessment carousel.

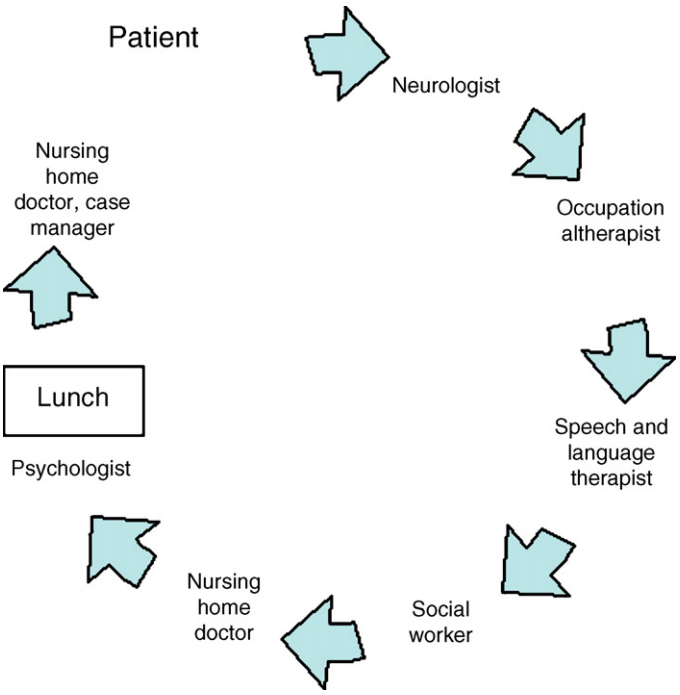


Fig. 1. Schematic representation of the activities during the visit to the out patient department.

3.3. Assessment tools

Each team member uses profession-specific questionnaires and testing material. The neurologist performs a general intake on neurological symptoms, inheritance and consequences from choreic movements and investigates the indication for symptomatic medication. The nursing home physician analyses somatic and functional problems as well as consequences of HD for the patient's social network. A cognitive screening is performed, using the Mini Mental State Exam (MMSE) [18]. The Behaviour Observation Scale Huntington (BOSH) [16] is completed with the caregiver and the Care Dependency Scale (CDS) [3] is filled in with the patient. The speech and language therapist tests the swallowing performance, dysarthria and language processing and production skills. The occupational therapist investigates mobility and functional capacity. She also observes activities of daily living (ADL) during lunch and manual dexterity for computer and/or communicator skills. To enhance

Author's personal copy

safety in and around the house, she visits the patient at home in the weeks following the outpatient department's first visit. The psychologist investigates mood, temper, flexibility and cognitive functions. The social worker analyses strong and weak parts of the social (supporting) system and inquires about leisure interests and activities during the day. Each patient's weight is measured in the clinic.

3.4. Establishing a care plan

After lunch, each professional verbalises the diagnosed problems and suggests treatment and goals in the multidisciplinary discussion. The combined observations result in an overall impression of the patient and how HD influences the patient's social system. The overall intention is to improve the quality of life of both patient and caregiver. This will enable the patient to function as a 'normal' family member and to stay home as long as possible and desirable. When the team decides another Huntington-specialised professional is needed for a particular problem (e.g. weight loss, mobility or behavioural problems), this can be easily arranged within the institute. The family physician and other medical doctors involved are informed through a copy of the care plan plus covering letter containing the diagnosed problems. Each professional has access to the care plan and can communicate with the case manager on changes or events that were not included in the plan. The case manager, a trained nurse with specialised skills in communication, is the linking pin in the whole care network. A sense of humour, a low threshold and perseverance are keywords for communicating with HD patients. If the patient or caregiver has a problem, he/she e-mails or calls the case manager who tries to solve the problem or mobilize a team member. As a principle local therapists strive for home treatment. The goals of this treatment are also included in the care plan. It requires a fine art of persuasion combined with flexibility and knowledge of the disease before the whole care plan starts rolling.

3.5. Home implementation plan

Part of a care plan is working on the patient's lifebook with the help of a social worker. This lifebook is a manuscript about the patients' life, habits, preferences, leisure interests and special dislikes. It may include pictures and special wishes or a declaration of intent. The advantage of making a lifebook with the patient and family is a 'healthy' approach to the patient. It is a pleasant way of communicating with a patient and it ensures the patient that the social worker is really interested in his/her patient. The patient is usually proud of his/her lifebook, which may also serve as a useful communication tool for the future if the patient is no longer able to speak or has to be admitted to a nursing home. The psychologist supports the patient and caregiver(s) in the weeks or months after the first visit. During this follow-up, a more elaborate explanation is given of the diagnosed cognitive changes and how to deal with it, as well as why overestimation and underestimation of the patient's capacities may lead to aggressive or apathetic behaviour. To prevent overestimation and to provide a meaningful daily living, one or more days of day care facilities are regularly advised. Although the majority of patients does not like this proposal at first, most of them really enjoy day care once experienced; it also enables their caregivers to go to work or have the day off. During day care, several therapies are offered. If indicated, a speech and language therapist advises and trains the patient and caregiver how to reduce choking risks and how to communicate optimally. If a communicating device may be necessary in the near future, education is being provided. If a specific part of the plan can be taken care of by a local therapist, this will be organized. To reach this goal, a lot of mutual arrangement is necessary. Local therapists, district nurses and day care staff are always trained how to treat an individual patient. The Huntington team together with the lay organization take care of these trainings. The local therapists can always fall back on the Huntington team from the outpatient department. To illustrate this coordinated multidisciplinary care, two case histories are given below.

4. Case histories

4.1. Case 1

A 58-year-old woman who was diagnosed with HD 6 years ago visited the outpatient department with her husband. They have two grown-up independent children. At the time, she had been suffering serious choking, for which her husband had to perform the 'Heimlich manoeuvre' six times. As a consequence, she could not eat on her own and her husband had to reduce his working hours. He also arranged for district nurses to accompany her during the meals, while he was working. Over the last years, she lost 5 kg of weight. She went to a day care facility twice a week. She did not want to go out on her own, because she recently fell and broke her pelvis. She smoked all around the house and sometimes forgot to put out her cigarette. She was short-tempered and tense and frequently talked about committing suicide. Her husband ceased the activities for his soccer club because he had to stay with her during the evenings and weekends. She appeared to have goitre. In the care plan a visit to the swallowing team in the local hospital was arranged to distinguish whether the choking risk was due to HD or to the enlarged thyroid gland. The test revealed that there was a complete inadequacy of swallowing due to HD and that a PEG catheter was indicated. She refused this and therefore all district nurses and day care workers were instructed to perform the Heimlich manoeuvre whenever needed. The psychologist also explained how to organize the meals in order to diminish the choking risk (e.g. no distraction, one thing at a time, and structure during the meal). The speech therapist together with the dietician explained and instructed the nurses how to supervise her and how to prepare the food and enrich it. The day care facility was arranged for 3 days instead of 2 and on the other weekdays accompaniment was arranged for. The physiotherapist went out walking with her and trained her how to relax and cough effectively. It was agreed upon that during her husband's absence she should only smoke in a separate room equipped with fire alarm. The occupational therapist advised a few adaptations in the house to increase safety during showering. Tandem-biking was trained. The social worker made a lifebook with the patient. During the second visit to the outpatient department her husband told that no Heimlich manoeuvre had been necessary during the last 6 months, that she had not fallen and had not mentioned any suicidal thoughts or plans. He returned to his fulltime job and resumed his leisure activities at the soccer club. The patient was really satisfied with both day care facilities and nursing care.

4.2. Case 2

A 39-year-old woman with two sons (3 and 7 years old) came with her partner to the outpatient department. She was the first patient in her family diagnosed with HD, about 6 months before. Later on, her father appeared to have the disease and her brother was identified as gene-carrier. She quit her job as chemical analyst about a year before because she started to make mistakes. Her colleagues advised her to visit a doctor and the diagnostic process started rolling. Her partner, a truck driver, felt he could no longer leave her alone with the children. She was apathetic and watched television all day long, went to bed late at night and could not get

Author's personal copy

started in the morning. In the afternoon she went to bed to sleep until about 4.30 p.m. thereafter she started cooking. When she was preparing a meal she could not keep an eye on the children. Sometimes, she had temper outbursts and left the house. Her partner had a fulltime job, so there was little time left to clean the house. His mother helped a lot by babysitting. A neighbour, who was a professional nurse, took care of the children most of the afternoons. The dysarthria of the patient and her decreased reaction speed impoverished communication with her children and her partner. In the care plan a district nursing team was instructed to assist the mother in her daily tasks like taking care of the children and being a housewife. The neighbour nurse was added to the team and together with explanation of the psychologist and the case manager and nursing home physician, the team started functioning. A specialised psychiatric nurse helped the nurses in the approach of the children's upbringing. Half a year later, the patient and her partner were contented with the way of living. The social worker made a lifebook with the patient and the patient trusted the team and could be persuaded to accept speech therapy on a weekly basis to improve communication. The family even bought a dog, which was a solace to both children and parents. The occupational therapist recommended a few adaptations in the bathroom to prevent falling on a slippery floor. To this moment, the patient still refuses this advice.

5. Closing remarks

Until now, care for HD patients, for whom nursing home care is not yet necessary, was not easily available and diverse in nature. Sometimes, patients visit a neurologist or a psychiatrist. Some patients have no care or treatment at all. In the literature on treatment of HD, medical professionals tend to strive for goals like quality of life, safety, mental well-being and optimal functional capacity. In this article a practical description is given of how this form of care is developed in the north of the Netherlands, in the setting of an outpatient department. The endpoints described in the relevant literature [1,8,14,9,10,2,15] fit the project aims. To make sure the care plan mentioned above is carried out well, a lot of communication between nurses, doctors, therapists and caregivers is necessary. The continuous backup from the Huntington team for families and local therapists appears time-consuming but inevitable for complete implementation of the plan. This can only be achieved by coordination. When the word "coordinate" is searched for in a dictionary the description is as follows: "bringing the different elements of a complex activity or organization into a relationship that will ensure efficiency or harmony". It is therefore that we suggest the term "coordinated multidisciplinary care" to be used for the treatment of HD as it is performed in our outpatient department.

This outpatient department has just started and has not yet proven its success. In the first project year 20 patients were treated. These patients, caregivers and professionals are positive about the care provided by the Huntington team. Questions that need to be addressed in the near future are the following: does the Huntington team's treatment enhance the patient's and/or caregiver's quality of life? And if so, how does it affect the disease process or behaviour of the patient? Another interesting topic is the need for residential care. Usually, the patient desires postponement of this need. But, if possible, does this also positively influence the quality of life of the partner or next of kin?

References

[1] A. Aubeeluck, E. Wilson, Huntington's disease. Part 1. Essential background and management, Br. J. Nurs. 17 (2008) 146–151.
[2] S. Dawson, L.J. Kristjanson, C.M. Toye, P. Flett, Living with Huntington's disease: need for supportive care, Nurs. Health Sci. 6 (2004) 123–130.
[3] A. Dijkstra, M. Coleman, T.W. Dassen, T.I. Romoren, M. Välimäki, R. Zanotti, Care dependency of nursing home patients with dementia: assessment from European perspective, Tijdschr. Gerontol. Geriatr. 31 (2000) 245–251.
[4] R. Gupta, P. Agarwal, J. Coates, Psychiatric expertise for a multidisciplinary Huntington's disease clinic ps.psychiatryonline.org/, 60 (2009) 267.
[5] K.F. Hoth, J.S. Paulsen, D.J. Moser, D. Tranel, L. Clark, A. Bechara, Patients with Huntington's disease have impaired awareness of cognitive, emotional and functional abilities, J. Clin. Exp. Neuropsychol. 29 (2007) 365–376.
[6] A. Kent, Huntington's disease, Nurs. Stand. 18 (2004) 45–51.
[7] J. Klager, A. Duckett, S. Sandler, C. Moskowitz, Huntington's disease: a caring approach to the end of life, Care Manage. J. 9 (2008) 75–81.
[8] M.L. Klimek, G. Rohs, L. Young, O. Suchowersky, M. Trew, Multidisciplinary approach to management of a hereditary neurodegenerative disorder: Huntington disease, Axone 19 (1997) 34–38.
[9] M. Nance, Comprehensive care in Huntington's disease: a physician's perspective, Brain Res. Bull. 72 (2007) 175–178.
[10] W. Phillips, K.M. Shannon, R.A. Barker, The current clinical management of Huntington's disease, Mov. Disord. 23 (2008) 1491–1504.
[11] R.A.C. Roos, A. Tibben, H.P.H. Kremer (red), De ziekte van Huntington. Begeleiding en behandeling, Elsevier gezondheidszorg, Maarsen, 2002.
[12] S. Simpson, Late stage care in Huntington's disease, Brain Res. Bull. 72 (2007) 179–181.
[13] S. Simpson, The management of Huntington's disease, Pract. Neurol. (2004) 204–211.
[14] H. Skirton, N. Glendinning, Using research to develop care for patients with Huntington's disease, Br. J. Nurs. 6 (1997) 83–90.
[15] B. Soltysiak, P. Gardiner, H. Skirton, Exploring supportive care for individuals affected by Huntington disease and their family caregivers in a community setting, J. Clin. Nurs. 17 (2008) 226–234.
[16] R. Timman, H. Claus, H. Slingerland, M. van der Schalk, S. Demeulenaere, R.A.C. Roos, A. Tibben, Nature and development of Huntington disease in a nursing home population. The Behavior Observation Scale Huntington (BOSH), Cog. Behav. Neurol. 18 (2005) 215–222.
[17] The Huntington's Disease Collaborative Research Group. A novel gene containing a trinucleotide repeat that is expanded and unstable on Huntington's disease chromosomes. Cell 1993 Mar 26; 72(6):971-83.
[18] T.N. Tombaugh, N.J.N.J. McIntyre, The mini-mental state examination: a comprehensive review, J. Am. Geriatr. Soc. 40 (1992) 922–935.
[19] E. van Duijn, E.M. Kingma, R. Timman, F.G. Zitman, R.A. Tibben, A. Roos, R.C. van der Mast, Cross-sectional study on prevalences of psychiatric disorders in mutation carriers of Huntington's disease compared with mutation-negative first-degree relatives, J. Clin. Psychiatry 69 (2008) 1804–1810.
[20] J.P. van Vugt, S. Siesling, K.K. Piet, A.H. Zwinderman, H.A. Middelkoop, J.J. van Hilten, R.A. Roos, Quantitative assessment of daytime motor activity provides a responsive measure of functional decline in patients with Huntington's disease, Mov. Disord. 16 (2001) 481–488.