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THE IMPACT OF HUNTINGTON'S DISEASE ON CAREGIVERS

The Czech experience

Por JAN ROTH*, TEREZA UHROVÁ**, PAVEL DOUBEK**, MAREK PREISS***,
JANA ZIDOVSKÁ****, EVA HAVRDOVÁ* y EVZEN RUZICKA*

RESUMEN

Objetivo: Evaluar los principales problemas en los cuales los cuidadores (CG) de pacientes con enfermedad de Huntington (HD) se ven enfrentados, desde los estadios tempranos de la enfermedad.

Diseño: 21 CGs (4 esposas, 8 esposos, 7 hijas, 1 madre y 1 hijo) fueron investigados en base a una entrevista estructurada con el uso de un cuestionario. Las áreas a evaluar fueron: A. Problemas en obtener información acerca de los estadios iniciales de la enfermedad. B. Problemas con el diagnóstico de la enfermedad. C. Comunicación con el médico. D. Manifestaciones clínicas que afectan a los CGs. E. Información acerca de grupos de apoyo así como la utilidad de éstos.

Resultados: 14/21 CGs tuvieron problemas severos en la obtención de información, 7/21 tuvieron información suficiente. El riesgo del desarrollo de HD en sus hijos fue el principal problema relacionado con el diagnóstico de la enfermedad. La comunicación con el médico tratante fue reportada como problemática en 15/21 CGs, mientras que en 6/21 la experiencia fue buena. Cambios afectivos y del comportamiento fueron reportados como los síntomas más severos de HD para 12/21 CGs (agresión en 12/21, depresión en 7/21 y problemas sexuales en 3/21), síntomas de demencia en 10/21, disartria en 7/21, movimientos involuntarios y/o desórdenes de la marcha en 3/21 CGs. La información acerca de la existencia de grupos de apoyo para HD entre médicos y programas de salud fue insuficiente para 16/21 CGs. La utilidad de los grupos de apoyo fue extremadamente importante para todos los CGs.

Conclusiones: Médicos y personal de salud deberían aceptar la idea de que los CG, aparentemente en buena condición física y de salud, necesitan tanto ayuda psicológica como médica.

* Movement Disorders Center, Department of Neurology, Katerinska 30, Prague 2, Czech Republic, Phone and fax No 004202 2496 5539, Email roth@beba.cesnet.cz.

** Department of Psychiatry.

*** Psychiatric Center Prague.

**** Department of Human Genetics, Charles University, Prague, Czech Republic.

ABSTRACT

Objectives: To evaluate main problems solved by caregivers (CG) of patients with Huntington's Disease (HD) from the onset of first symptoms to advanced stages of HD.

Design: 21 CGs of HD patients (4 wives, 8 husbands, 7 daughters, 1 mother, and 1 son) were investigated by means of structured interview using the questionnaire. Following main topics were evaluated: a. problems getting adequate information about HD in early stages of the disease, b. main problems connected with the diagnosis itself, c. cooperation and communication with physicians, d. clinical symptoms of HD most severely affecting the life of CG, e. information about the existence of HD support group and the usefulness of HD support group for CG.

Results: 14/21 CGs had severe problems with getting adequate information about HD in the early stage of HD, while 7/21 had enough information. The risk of HD for their children was found to be the main problem connected with the diagnosis itself. Cooperation and communication with physicians were reported as very problematic in 15/21 CGs, whereas 6/21 had good experience with physicians. The insufficient interest in family and social problems and almost no contact with partners and CGs were reported to be the main controversy. Behavioral and affective changes were reported as most severe symptoms of HD for 12/21 CGs (aggression in 12/21, depression in 7/21 and sexual disturbances in 3/21), symptoms of dementia in 10/21, dysarthria in 7/21, involuntary movements and/or gait disorders in 3/21 CGs. The information about the existence of HD support group among physicians and health care services was found to be insufficient in 16/21 CGs. The usefulness of HD support group was found to be extremely important for all CGs.

Conclusions: All physicians and medical staff should accept the idea that CGs, apparently healthy persons, in fact need psychosocial as well as medical help.

PALABRAS-CLAVE : Enfermedad de Huntington, cuidador, grupos de apoyo.

KEY WORDS : Huntington's Disease, caregiver, support group.

INTRODUCTION

Huntington's disease (HD) is an autosomal dominant inherited progressive neurodegenerative illness. Most of the time, personality and behavior disorders, intellectual deterioration and involuntary movements manifest the disease. Due to its complexity, HD is a typical example of the disease that affects not only its carriers but also the whole family. The principal direct impact of the disease is its mercilessly progressive character and the 50% risk of the transmission on the offspring.

The main indirect impact is the stress connected with the nursing of the sufferers, decisions on the predictive tests for HD at-risk people and a lowered socioeconomic status of the family caused by the disability of the HD patient.

The HD research is focused mainly on the biological principle of the disease and on search for a possible therapy. A great attention is given to HD at-risk persons.

However, problems and needs of caregivers (CG) nursing the patient are not in the center of the attention¹⁻⁵.

Thus, the goal of our work was to find the priority problems and needs of CG and find out how to help and support them.

METHOD

21 CG of our HD support group (4 wives, 8 husbands, 7 daughters, 1 son and 1 mother) participated in this study, using a structured questionnaire. Every interview took 60 minutes. All but one HD patient had children. The HD predictive test was performed only in one CG (the son).

The following five topics were evaluated:

- a. Problems with getting adequate information about HD in early stages of the disease.
- b. Main problems with the diagnosis itself.
- c. Physician - CG cooperation and communication.
- d. Clinical symptoms most severely affecting CG – suffering person coexistence.
- e. Information about HD support group and the usefulness of the group for CGs.

RESULTS

a. Problems with getting adequate information about HD in early stages of the disease

In early stages of the disease, 14 out of 21 CG (66%) had serious problems with getting information about HD from attending physicians, only 7 out of 21 (33%) got sufficient information.

b. Main problems with the diagnosis itself

Every CG reacted to the diagnosis with a fear from the incurable hereditary disease.

The main concern of all spouses was the risk of the genetic transmission to their children. The incurability of the illness was named as the second problem. Children as CG answered less exactly and talked about common fear after learning the parent's diagnosis.

c. Physician - CG cooperation and communication

Physician - CG cooperation and communication was evaluated as bad and insufficient by 15 out of 21 CGs (71%); 6 out of 21 CGs (29%) had a good experience with physicians. The shortage of interest in the family and the social problems of CGs and almost no contact with CGs principally caused the dissatisfaction with physicians.

d. Clinical symptoms most severely affecting CG – suffering person coexistence (maximum 3 symptoms)

Behavior and affective changes were described by 12 out of 21 CGs as the most serious symptoms of the disease (9 cases of verbal and physical aggression (7 female CGs, 2 male CGs), 7 cases of depression, 3 cases of sexual violence on CGs - wives). Dementia symptoms were found as the most severe by 10 out of 21 CGs, dysarthria by 7 out of 21 CGs, involuntary movements and/or gait disorders by 3 out of 21 CGs (see in Table 1).

e. Information about HD support group and the usefulness of the group for CGs

Sixteen out of 21 CG (75%) did not get any information about HD support group from the attending physicians but learned about it later, often during genetic counseling or consulting a physician cooperating with the group. All CGs evaluated highly the

activities of HD support group such as giving information and support for their future life and help to the HD affected persons.

DISCUSSION

Our study leads us to formulate the following ideas:

- a) The problems with getting the adequate information about HD in early stages of the disease may result from the fact that general practitioners and regional neurologists are not aware of this relatively rare illness. This may indicate that physicians are neither informed sufficiently about the possible further investigation and care at movement disorders centers nor aware of the existence of HD support group. The physicians may even lack courage to confront the problem and spread the serious information that would threaten the whole family.
- b) The main problem of the CGs – spouses is the fear that the disease would break out in their children. CGs accept their spouses disease less complicatedly than the risk for their offspring. For CGs, their feelings of powerlessness (in the incurability of the disease, their insufficiency to handle the situation, the risk of the disease for their offspring), anger (at the ill person, bad fate, the uninterested environment) and guilt (of outbursts of rage in front of the sufferer, of the possible disease of their children) were characteristic. The diagnosis was concealed from adult children – HD at-risk person in one family and from a future daughter's spouse – HD at-risk person. It is questionable whether physicians themselves should seek for further relatives and inform them about the risk as the interests of the sufferer, CG and a HD at-risk person might interfere. The CGs – offspring of HD patients could express a common fear only and be ashamed to be worried about their own fate. It is necessary to point out that 38% of CGs in our study are HD at-risk persons.
- c) The physicians seem to cooperate with CGs unsatisfactorily. HD obtained the reputation of a completely incurable illness and a total fatalistic approach has prevailed. Although many HD psychosocial problems could be solved with the CG help, CGs had problems that attending physicians neither communicated, had appointments with them nor showed interest in their nursing problems. On the other hand, the powerlessness of the physician in the HD prognosis might be interpreted by CGs as the ignorance and lack of information. Therefore, the team of physicians dealing specifically with HD was established in the Czech Republic as late as several years ago.
- d) Clinical symptoms most severely affecting CG – suffering person coexistence were changes of the mental state such as changes of behavior, personality and emotions. We were able to recognize three cases requesting the acute psychiatric intervention of CGs (deep depression, repeated rape). The adequate care has not been given in any case. However, no obvious request for help was indicated. Movement disorders were not found to be the most tragic symptoms of HD (obviously, CGs were informed about them thoroughly by physicians and from literature). Probably, the symptoms they did not expect became main problems for CGs.
- e) Information about HD support group was not satisfactory accessible. Four patients learned about the existence of the group from a geneticist, a HD specialist and the

president of the group in one person. As the support group was established in the Czech Republic, as early as in 1991, there was time enough to spread the information about its existence. HD support group has its web page and the journal that has been distributed to geriatric institutes and psychiatric hospitals and later spread among neurologists, too. However, our observation is retrospective and we do hope, the information is nowadays more accessible for physicians and HD families.

All CGs found HD support group as extremely beneficial for getting adequate information about HD and its inheritance, presymptomatic and prenatal testing, socializing with people of a similar fate and exchanging of experience and information about particular problems.

CONCLUSION

The obtained results are in many points alarming and they require amelioration of the present approach to CGs. Some facts we mention possibly reflect the situation in other countries.

We think that these particular steps should be taken:

1. All physicians and medical staff should accept the idea that CG, apparently healthy persons, in fact need psychosocial as well as medical help.
2. It is necessary to spread the knowledge about the complex topic of HD and CGs in the large professional public.
3. It is necessary to have the easy possibility to send the patients to movement disorders centers and to enable broad access to the HD support group.
4. It is also necessary to participate actively in the family and social support and to collaborate more intensively with social workers.

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TABLE 1

CLINICAL SYMPTOMS MOST SEVERELY
AFFECTING CG – SUFFERING PERSON COEXISTENCE
(maximum 3 symptoms).

The principal symptom	CG number (n-21)	%
Behavioral changes	12	57
Dementia	10	48
Aggression	9	43
Depression	7	33
Dysarthria	7	33
Involuntary movement	3	14
Gait disorders	3	14
Sexual violence	3	14