ABSTRACT

Objectives: to report the application of the Systematic Nursing Assistance to patients with Huntington Disease (HD). Method: this was a descriptive study with a qualitative approach, in the format of an experimental report. Assistance to HD patients and their families were provided between April 3 and 30 of 2011 in the Internal Medicine ward of the Maria Aparecida Pedrossian/NHU-UFMS University Hospital, in Campo Grande/MS/Brazil. This study was approved by the Ethics in Research Committee from the Federal University of Mato Grosso do Sul under the protocol number CAAE 0340.0.0.049.000-11. Results: the Systematic Nursing Assistance to patients with low prevalence diseases, or diseases that are not often seen in hospital institutions favors individual and systematic visions of the person affected by the disease. Conclusion: there are few studies related to these patients, especially about nursing care, indicating the need for studies involving these patients. Descriptors: Huntington Disease; Nursing Processes; Nursing Care.

RESUMO

Objetivo: relatar a experiência da aplicação da Sistematização da Assistência de Enfermagem ao paciente com a Doença de Huntington (DH). Método: estudo descritivo, de abordagem qualitativa, do tipo relato de experiência. Realizou-se a assistência ao paciente com a DH e a família, no período de 3 a 30 de abril de 2011, na enfermaria de Clínica médica do Hospital Universitário Maria Aparecida Pedrossian/NHU-UFMS, em Campo Grande/MS/Brazil. Este estudo teve o projeto de pesquisa aprovado pelo Comitê de Ética em Pesquisa, CAAE 0340.0.0.049.000-11. Resultados: a Sistematização da Assistência de Enfermagem à pacientes com doenças de baixa prevalência ou que não são atendidas frequentemente em instituições hospitalares favorece a visão individual e sistemática da pessoa acometida pelo agravo. Conclusão: existem poucos estudos relacionados a estes pacientes, principalmente em relação à assistência de enfermagem, gerando a necessidade estudos que os envolvam. Descritores: Doença de Huntington; Processos de Enfermagem; Cuidados de Enfermagem.

RESUMEN

Objetivos: informar la aplicación de la Asistencia Sistématica de Enfermería a pacientes con la enfermedad de Huntington (EH). Método: es un estudio descriptivo con un enfoque cualitativo, en el formato de un informe experimental. Los pacientes con EH y sus familias recibieron asistencia entre 3 y 30 de abril de 2011 en la sala de medicina interna del Hospital Universitario de Maria Aparecida Pedrossian/NHU-UPM, en Campo Grande, MS, Brasil. Este estudio fue aprobado por la Comisión en la Ética de Investigación de la Universidad Federal do Mato Grosso do Sul bajo el número de protocolo CAAE 0340.0.0.049.000-11. Resultados: la Asistencia Sistématica de Enfermería a pacientes con enfermedades de baja prevalencia o enfermedades que no se ven a menudo en las instituciones hospitalarias favorece visiones individuales y sistemáticas de la persona afectada por la enfermedad. Conclusión: son pocos los estudios relacionados con estos pacientes, especialmente sobre el cuidado de enfermería, indicando la necesidad de estudios en estos pacientes. Descriptores: Enfermedad de Huntington; Procesos de Enfermería; Cuidados de Enfermería.

1Resident nurse, Multi-professional in Attention and Care to Critical Patients Residence Program/NHU/UFMS. Campo Grande (MS), Brazil. E-mail: ccsar.msf@hotmail.com; 2Resident nurse, Multi-professional in Attention and Care to Critical Patients Residence Program/NHU/UFMS. Campo Grande (MS), Brazil. E-mail: mdfcheade@uol.com.br; 3Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 4Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 5Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 6Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 7Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 8Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 9Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 10Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 11Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 12Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 13Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 14Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 15Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 16Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 17Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 18Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 19Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 20Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 21Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 22Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 23Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 24Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 25Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 26Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 27Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 28Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: gorettereis@gmail.com; 29Nurse, Professor, Doctor, Nursing School, Federal University of Mato Grosso do Sul. Tutor at the Multi-professional in Attention and Care to Critical Patients Residence Program /NHU/UFMS. Campo Grande (MS), Brazil. E-mail: goren```
INTRODUCTION

Huntington Disease (HD), described by the physician George Huntington in 1872, consists of a neurodegenerative disturbance, also known as hereditary Chorea. 1 This disease presents an autosomal dominant pattern featuring neurologic, autonomic, and neuropsychiatric manifestations. 2

The HD is present worldwide, however, there are reports of high incidence of this disease in some regions such as Maracaibo, in Venezuela. 3 There are no epidemiological studies that show its statistics in Brazil, however, its incidence is estimated as 5 to 10 cases per 100,000 inhabitants; there are no predisposing factors described for its occurrence. 1

There are entities that support patients affected by Huntington disease and their families in Brazil. The Huntington Brazil Association (ABH) and the Union of Relatives and Friends of HD patients (UPADH), with headquarters in Sào Paulo and Brasilia, respectively, 3 are examples of sites that offer assistance to these patients. There are no reports indicating genre biased prevalence; HD equally affects men and women. Higher prevalence in Caucasians have been reported, however, it occurs in all ethnic groups. 4

Because it is a progressive degenerative disease of the nervous system, with autosomal dominant inheritance pattern and complete penetrance, all individuals who carry the DH genotype will, at some point in their lives, present signs and symptoms of the disease. 1 HD individuals present destruction of striated cerebral nuclei neurons whose function is the production of the GABA neurotransmitter. This episode determines the appearance of irregular and involuntary movements and progressive mental decline caused by the reduction in GABA production in their nervous system. 1

HD is transmitted through the autosomal dominant IT15 gene located on the short arm of chromosome 4 (4p16.3). This gene shows an excessive number of CAG repeats (cytosine-adenosine-guanine), which is considered abnormal when present beyond 40 repeats because the normal number of repeats is between 9 and 34. 3, 5 In addition, this gene encodes the huntingtin protein that presents an abnormal amount of glutamine amino acids in its chain, which leads to neuronal dysfunction, generalized brain atrophy, changes in neuroreceptors levels, and accumulation of neuronal and cytoplasmic proteins aggregates. 1

OBJECTIVES

- To report the application of the Systematic Nursing Assistance (SAE) to patients with Huntington Disease (HD)
- To discuss the peculiarities of families with Huntington disease (HD).

METHOD

This was a descriptive study with a qualitative approach, in the format of an experimental report. Assistance to HD patients and their families were provided during patients’ hospitalization, between April 3 and 30 of 2011, in the Internal Medicine ward of the Maria Aparecida Pedrossian/NHU-UFMS University Hospital, in Campo Grande/MS/Brazil.

The service was conducted by a multi-professional team of nurses, physical therapists, nutritionists, pharmacists, and a dentist surgeon. These professionals are part of the Multi-professional in Attention and Care to Critical Patients Residence Program/NHU/UFMS in this institution.

The patient was followed by the team and each case was discussed at a weekly clinical
discussion between professionals in the multi-professional residence program and preceptors in the medical areas that compose the program.

Through SAE guidelines, the nurse evaluated the patient’s medical history, composed by anamnesis and physical examination, listed the nursing diagnoses, and defined treatment goals and nursing interventions. The NANDA Taxonomy System (North American Nursing Diagnosis Association) was used in this study to carry out SAE evaluation. The publication of the data was authorized by the family members given the cognitive deficit of the patient. This authorization was obtained through their signature in a Voluntary Informed Consent (TCLE). This study was approved by the Ethics in Research Committee from the Federal University of Mato Grosso do Sul under the protocol number 2255 CAAE 0340.0.0.049.000-11.

RESULTS

Nursing-history - Male patient, 33 years old, white, evangelical, incomplete higher education, born in Campo Grande-MS, where he currently resides with his mother, four brothers, and a nephew. The family monthly income is of eight minimum wages. Pensioner, bedridden, and without an active life before becoming an inpatient (after onset of symptoms). Denies smoking and alcoholism history. Symptoms started about five years ago, however, the disease worsened two years ago, when the patient became bedridden and non-cognizant. The patient had an active life, regular physical activity, and was studying Civil Engineering before the onset of symptoms. The manifestation of symptoms was gradual and began after the death of his father. There is a family history of Huntington disease in cousins, an aunt, and a brother who is a few years older. The father, grandfather, and older brother presented similar symptoms, however, with greater impact towards depression. The patient was diagnosed with Huntington disease eight years ago.

Physical Examination - Bedridden, conscious, non-cognizant, with periods of psychomotor agitation, and choreic movements. Pale ocular mucosa 2+/4+, normal sclera, isochoric and photo reagent pupils; pale oral mucosa 2+/4+. Symmetric thorax, good expansibility, lungs sound clear to percussion in all areas. Physiological vesicular murmurs present, however, diminished in both bases. Normophonetic and rhythmic heart beats in two tempo without

blows. Semi-globus and flabby abdomen, RHA (+), tympanic sound to percussion. Painless to superficial and deep palpation. Peripheral perfusion present (3s), strong, symmetrical, and rhythmic pulses. Edema +++/4+ MMSS and MMII. Nasogastric tube feeding (SNG), 5 times a day (1,000 ml - 1200 Kcal). Bowel movements present with pasty consistency, brownish, and with vesicle eliminations through an urinary catheter (SVD), of citrus yellow color and without sediment. Invasive Devices: SVD, SNG, peripheral venous access (AVP), hemodialysis catheter in the left subclavian. Applied Scales: FUGULIN: Semi-intensive Care; BRADEN (15 - moderate risk) GLAGOW: 12 (AO: 4-RV: 2-RM: 6).

Neurological examination - Currently presenting involuntary movements with active attitude. Choreic faces (exaggeration and non-opportunistic expressions) conscious, lethargic, and no sense of time and space.

Balance, gait, and coordination were not evaluated because the patient is bedridden and disoriented. Tropism and eutrophic state alterations, and muscle group hypotrophy were not observed. Present MMII and MMSS low grade hypertonia. Sustains muscle strength at grade 5 in all muscle groups. Preserved deep reflexes, Babinski’s positive sign. Presents preserved protopathic superﬁcial sensitivity. Insufficient evaluation of epicritic sensitivity. The evaluation of deep sensitivity and stereognosis were also not possible. The evaluation of dermatomes and pairs of cranial nerves was only possible on the II and III pairs, optical and oculomotor, respectively. The response was normal on these, with isochoric and photo reagent pupils and eyeball movements in all directions.

Nursing diagnoses and care procedures during hospitalization.

Risk of infection (primary inadequate defenses, increased environmental exposure to pathogens, invasive procedures).

- To monitor sites with insertion of invasive devices;
- To perform dressing in double lumen catheter insertion sites for hemodialysis;
- To guide escorts and visitors about infection control measures.

- Impaired physical mobility related to medicines, cognitive and musculoskeletal impairment characterized by non-coordinated movements.
- To perform decubitus changes;
- To apply the BRADEN scale daily;
- To apply the BRADEN scale daily;
- To install air mattress;
- To use cushions for comfort;
Ferraz CCB, Ortega FB, Reis MG dos et al.

- To emulsify skin and bony prominences.
  ✓ Aspiration hazard (feeding tube, impaired swallowing).
- To keep the head of the bed elevated to 30° continuously;
- To guide diet administration.
  ✓ Deficit in self-care for bathing related to cognitive impairment characterized by the inability to wash the body.
- To perform bed bath;
- To perform oral hygiene
  ✓ Deficit in self-feeding related to cognitive impairment characterized by inability to ingest food safely.
- To elevate bed head in 45°;
- To guide nasogastric tube care;
- To administer diet by nasogastric tube;
  ✓ Risk of traumas (cognitive difficulties, history of previous trauma).
- To keep high bed grills;
- To use containment devices in bed when necessary;
  ✓ Caregiver role strains related to 24 hours a day of caring responsibilities, concern with respect to family members characterized by social life distancing and stress.
- To seek meaningful person/support network;
- To request social evaluation;
  ✓ Impaired social interaction related to communication barriers and disturbed thought processes characterized by dysfunctional interaction with other people.
- To seek meaningful person;
- To maintain therapeutic communication.

Other diagnostics that contribute to the perception of problems related to hospitalization of the patient were assessed in addition to the cited nursing diagnoses, including: Impaired skin integrity; Excessive liquid Volume; Hyperthermia; Risk of electrolyte imbalance; Risk of ineffective renal perfusion; Risk of falls; and Impaired verbal communication, among others.

**DISCUSSION**

The patient and their family members received nursing, multi-professional, and hospital staff assistance during approximately 30 days.

The Systematic Nursing Assistance to patients with low prevalence diseases, or diseases that are not often seen in hospital institutions, favors the individual and systematic visions of the person affected by the disease. This provides an integral care to the individual and their families.7

The use of SAE as the base of offered assistance made the organization of thoughts, observation, and intervention of nurses possible. It also became a systematic and logic structure toward actions related to the patient’s health in the hospital environment, directing the solutions of problems and providing criteria to evaluate the effectiveness of these procedures.

The survey of the nursing diagnoses indicated that, apart from the characterization of a patient with a high degree of complexity, high dependence on family, and hospital nursing care, there are problems related to psychosocial segments, which are common in families with people under these conditions who need constant care and attention.

We must remember that the nurse must accommodate the caregiver during nursing procedures and give him subsidies during the patient’s hospitalization. Thus, the caregiver will be more prepared and confident to care for their family member at home. We must also consider a network to support this caregiver, whether through family members, religious groups, or associations of specific groups.

The staff was present, during hospitalization, in the preparation of family members for the patient’s hospital discharge. The nurse could plan the assistance through SAE, identifying priorities and establishing actions to be taken by the nursing staff and multi-professional residency team for short-, medium-, and long-term care.

**CONCLUSION**

Huntington disease has a chronic-degenerative character, making the affected individual dependent on their family, which creates stress and changes in the life of everyone involved. Because this is a disease of low incidence, it is not discussed during the training of nursing professionals. However, the social repercussions for people affected by this disease generate the need for multi-professional care to improve the quality of life of all involved. Nursing care that, during hospitalizations, is dedicated to prepare the family and patient for a successful hospital discharge and lower the risk of re-hospitalization is of great importance because this condition of dependency will follow the patient who will not have a prognosis for self-care or chances to improve cognitive aspects.
Nursing care must be prepared to meet the patient’s and their families’ psychosocial needs. There are few studies related to these patients, especially addressing nursing care, which indicates the need for studies involving these patients. This study contributed to the experience in comprehensive care to patients affected by Huntington disease and their family members.

REFERENCES


