Care management of a patient with Devic’s Disease in Primary Health Care*

Objective: Describing the care management of a patient affected by Devic’s Disease in the Primary Health Care setting. Method: A clinical-qualitative case study based on the health status of a Devic’s Disease patient in a Health Center of the municipality of Florianópolis, accompanied by the Family Health Team. Data collection was carried out by electronic medical records, documents of the patient’s domain, as well as a semi-structured interview with the participant. Ethical aspects of research involving human beings were respected. Results: The categories were defined according to relevance criteria with the purpose of reporting the case study, presenting a unique implemented therapeutic project and describing the patient’s perception of her situation. Conclusion: Acupuncture and auriculotherapy were successfully performed, evidencing an improvement in the patient’s pain, which may contribute to new possibilities of care. Despite this, not all the available care tools were implemented, considering the range of complementary care therapies that go beyond the medicinal approach.

ABSTRACT

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Descriptors

Rare Diseases; Neuromyelitis Optica; Primary Health Care; Primary Care Nursing; Comprehensive Health Care; Complementary Therapies.


1 Universidade Federal de Santa Catarina, Centro de Ciências da Saúde, Programa de Pós-Graduação em Enfermagem, Florianópolis, SC, Brazil.
2 Secretária Municipal de Saúde de Florianópolis, Florianópolis, SC, Brazil.
3 Universidade Federal de Santa Catarina, Centro de Ciências da Saúde, Florianópolis, SC, Brazil.
INTRODUCTION

In 1894, the French physician Eugene Devic described a case report of subacute myelitis with severe bilateral amaurosis for the first time, and in his honor the pathology is described as Devic’s Disease or Neuromyelitis Optica (NMO). For standardization purposes we will use the term Devic’s Disease in this study.

Devic’s Disease is among rare pathologies which are characterized by affecting up to 65 people per 100,000 individuals, with a prevalence of 1 to 4.4 per 100,000 people in the Western world[2-3]. It is included in the group of Idiopathic Inflammatory-demyelinating Diseases (IIDDs), immunomediated and necrotizing the Central Nervous System (CNS), which gather acute or chronic clinical conditions of unknown etiology characterized by neurological manifestations, inflammatory changes, clinical evolution and severity, and its characteristics include involvement of the optic nerve and the spinal cord(1).

This disease predominantly affects females in comparison to males, with a mean age of 39 years at symptom onset. However, cases in childhood and in the elderly have also been described. It may present relapses in 80 to 90% of cases or be monophasic in 10 to 20%, frequently associated with other autoimmune diseases such as Myasthenia Gravis, Systemic Lupus Erythematosus, Sjögren’s Syndrome, Celiac Disease, and Sarcoidosis. Moreover, 20 to 30% of the cases are preceded by infection or vaccination(3).

Despite being affected at different severity levels, patients with this disease, including relapses in some cases, are not always hospitalized or do not always require tertiary care from the Health Care Network (HCN). Thus, we can highlight the importance of care management in the Primary Health Care (PHC) as being responsible for the follow-up and continuity of care.

Care management is related to the management of care networks, the articulation of resources and the development of strategies to ensure the principle of comprehensiveness in health care. It is also based on the organization and interaction among professionals at the (different) care levels of the health system, contributing to an expanded clinical approach and the qualification of health care(4-9).

The care of rare diseases under the PHC and the HCN is described in the Ministerial Ordinance Number 199, dated January 30, 2014 of the Ministry of Health (MoH), which establishes the National Policy for Comprehensive Care for Persons with Rare Diseases and approves the Guidelines for Comprehensive Care for Persons with Rare Diseases within the scope of the Brazilian Unified Health System (SUS – Sistema Único de Saúde), with the objective of reducing morbidity and mortality and secondary manifestations, seeking to improve the quality of life of these people through actions for promotion, prevention, early detection, timely treatment, reduction of disability and care(2).

This Ordinance defines the structure of care to people with rare diseases, and the PHC is responsible for coordinating care, since it provides continuous attention to the population, even when patients are referred to other HCN care services. In addition, it must guarantee counter-referal from specialized services to PHC, seeking to continue clinical follow-up and network support(2).

In this way, PHC is the preferred contact point for users, the main gateway and the HCN coordination center for care. It aims for an increase of access and connectivity, equity, universality and comprehensiveness in health care(6). This structure can be evidenced in the city of Florianópolis, which has a Health Care Network organized in such a way that the PHC coordinates the care of individuals. It is worth emphasizing that Florianópolis has been named as the Brazilian capital with the best PHC services, with 100% Family Health Strategy coverage in 2015, according to data from the Department of Primary Care of the Ministry of Health(7).

Based on the this scenario, this study is justified by the need to qualify clinical practice and health professionals’ knowledge, especially nurses, regarding the comprehensiveness of care to individuals with rare diseases and/or other patients with degenerative and demyelinating diseases. From the scientific point of view, this study is relevant due to the scarce production of knowledge regarding the care of patients affected by Devic’s Disease in the PHC, with a comprehensive focus on the individual and the use of integrative and complementary practices for handling their cases. In both Brazilian and international literature, studies on Devic’s Disease focus on case reports and the description of hospital-centered care.

Thus, this study aimed to describe the care management of an individual patient affected by Devic’s Disease in the PHC context.

METHOD

This is a clinical-qualitative case study in which the sample consisted of a variety of types according to the patient’s health state(6). This methodology is used to understand the meaning attributed by the patient to the health-disease process in the context where this phenomenon occurs, meaning the healthcare setting(9). Thus, the focus of the study is the subject’s statements regarding her health and illness process, the health services used, the therapeutic processes performed and the essence of her experience regarding the subject(8).

In this way, a Health Center of the municipality of Florianópolis was used as the research scenario to report the case of a patient affected by Devic’s Disease followed by the reference FHS (Family Health Strategy) of the region. The patient was approached in person at the Health Center during one of her consultations and invited to participate in the study.

Data collection was performed in the period from August to September 2016 using data from the electronic medical record (Infosaúde), patient documents and a semi-structured interview with the research participant.

In the documentary analysis, the electronic medical record and the documents made available by the patient such as reviews of hospital discharge, examinations and prescriptions provided the information on the care given at the PHC and at other health care levels.
The patient affected by Devic’s Disease is female, age 35, having brown skin, married, and who resides with her husband and daughter. She worked as a secretary at a private school. She is originally from São Paulo–SP, but she had been residing in Florianópolis–SC for more than 5 years, being attended by the reference FHS at a Health Center in the municipality.

Prior to her illness, she wore glasses and suffered from sporadic migraines. She denied having any comorbidities or using continuous medications. The case began in March 2010 when she used the PHC as the gateway with complaints of frontotemporal hemicranial headaches associated with pain, right amaurosis and loss of color.

Due to an exacerbation of the symptoms she sought care from an ophthalmologist, and according to the performed evaluation there were no alterations in the fundus of the eye or ophthalmological alterations. She was referred to the neurology service of the city hospital, reaching the Tertiary level of Care.

Complementary Diagnostic Tests (CDT) were then performed at the hospital to investigate possible pathologies. A lumbar puncture with CSF (cerebrospinal fluid) showed no significant changes and no alterations were identified by Computed Tomography (CT scan) of the skull or by Magnetic Resonance Imaging (MRI). Thus, the patient underwent corticosteroid treatment for 5 days, presenting improvement in the symptoms.

In January 2011, 1 year after the first crises, she underwent an anti-neoinrmyleite antibody test (aquaporin 4), which was reactive: 1/160. This is an important marker for the disease, enabling accurate pathology diagnosis.

In the year 2011, the patient presented three acute attacks of the disease. After the first crisis, an interval of 1 month preceded the second crisis, and another interval of 3 months preceded the third. In addition, the symptoms during the attacks coincided: headache in the frontal and right supraorbital regions, and reduced visual acuity in the right eye with significant visual impairment. During this period, she underwent a new skull MRI, which was within the limits of normality, an examination of the fundus of the eye showed no alterations, and there were no other signs of relevance to the physical examination. She underwent pulse corticosteroid therapy for all crises, and an increase of twice the medication dose was evidenced in one of them.

Up to this point the patient was using the PHC as the gateway; first when the symptoms started, and sporadically for renewing her prescriptions. She continued her follow-up in the Tertiary Care with a neurology team from a city hospital, conducting periodic check-ups.

After almost 2 years without crisis she had two occurrences in 2013, with more severe manifestations of the disease involving (in addition to headache) pain at touching the scalp, pain in the right eye, neck pain, low back pain and paresthesia in the lower left limb (LLL). She underwent a lumbosacral spine radiography, which showed a reduction of the L5-S1 space, and radiography in the hip which did not evidence any structural alterations. She underwent pulse corticosteroid therapy with Beta Trinta® replacement, demonstrating improvement in inflammatory and pain symptoms.
In the following year (2014), three crises occurred again with the same described symptoms of headache, retro-ocular pain, reduced visual acuity in the right eye with loss of color vision and blurred vision, difficulty reading, and difficulty recognizing figures. A fundoscopic examination showed an increase in the excavation and signs of optic atrophy in the right eye, while the left eye was within normal range and there was no prognosis for improvement in visual acuity. In addition to the signs and symptoms that affected the optic nerve, symptoms related to the spine were also evidenced, which were: paraesthesia in the Upper Left Limb (ULL) and in the LLL, difficulty in locomotion and gait, and reduced strength in the LLL.

In 2015, the MRI examination of the skull revealed signs of optic atrophy on the right side and reduced thickness. An MRI of the cervical and dorsal spine showed an extensive lesion with demyelinating characteristics compromising the cervical segment of the spinal cord from the medullary level to C6, with no signs of inflammatory activity. A degeneration of cervical intervertebral discs and small posterior disc protrusions at C3 to C7 levels, more pronounced at C5-C6 were also identified.

**Development of a unique therapeutic project for care management**

The description of the case pointed out the shared care between primary care and other levels of care. However, at a certain point the care was centralized at the hospital level, with a focus on medical treatment and the use of care technologies focused on drug therapy.

Based on the complaints reported by the patient who presented symptoms related to pain, a Unique Therapeutic Project aimed at the care of the patient was developed by the FHS in early 2016, with shared care decision-making between the FHS and the patient. On this occasion, the inclusion of Integrative and Complementary Practices (ICP) from Traditional Chinese Medicine (TCM) were proposed in addition to conventional medical treatment carried out by Biomedicine; more specifically acupuncture and auriculotherapy with weekly treatment sessions performed by physician and nurse\(^2\), respectively. After 1 year of complementary treatment with 17 acupuncture sessions and four auriculotherapy sessions, a stabilization of the condition was observed. No new acute attacks of the disease were reported. Similarly, the FHS continued to monitor and care for the case.

**The patient’s perception of her health status**

This category presents the patient’s perception regarding her health situation, as well as the diagnosis and treatment stages of the disease.

The patient initially described the flows and referrals throughout the treatment, the waiting time for receiving care and for obtaining information about her health condition, according to the following speech:

_I was referred to the hospital with a referral letter for the neurologist, and I stayed there all day crying, suffering, then I was introduced to a doctor who thought I had multiple sclerosis (...) At this point I was very scared, I was very afraid of multiple sclerosis (P)._  

The patient reported the emotions and afflictions present in the course of the disease, mainly in the discovery (of the diagnosis), which she reported as: frightened, doubtful, anguish, and fear: _I thought I was going to die._ (P).

These emotions were exacerbated by the fact that it was a rare and little known disease, even by health professionals. Thus, she reported the discovery moment of the disease: _They asked me to do an antibody test called aquaporin 4, so that they could be sure whether or not I had multiple sclerosis or what I had. And from that exam I was diagnosed with Devic’s Disease (P)._

And what followed after the diagnosis: _That was when I started to be aware of what I really had, that it was a disease that was going to have relapses, which could happen in the future affect my spine, my cervical, as it did last year, I could lose movements, speech, have difficulty breathing, all that. So, that was when I began to understand more or less what was happening to me, and I thought I was not going to survive (P)._  

Another point highlighted by the patient were the crises or acute phases of the illness, which occurred during periods of stress in her personal life, a triggering factor for the onset of the outbreaks. According to the patient’s perception, stress is what precedes every crisis of the disease. Consequently, worsening of the disease occurred with greater frequency in more troubled years.

Furthermore, the pain was representative according to the patient’s perception. This symptom was mentioned more than once during the interview, with the patient characterizing it as: _an unbearable pain_ (P), and remembers about the waiting time for care while feeling a lot of pain, in addition to reflecting on the complexity of measuring somebody’s pain: _we never know the extent of another person’s pain_ (P).

The speeches emphasize the treatment associated with Integrative and Complementary Practices as beneficial and very important for her recovery: _Acupuncture was the finding of my life, It’s true. It was something that lifted me up when I was practically on the ground, okay. When I was discharged from the hospital I did not have any options (...) So I was looking for something to get me out of that situation I was in. So when I came here, and I experienced acupuncture, it was a kind of liberation from my life, it helped a lot. I think this should always continue at the health clinic, they should always offer it because it really helps (P)._  

The patient continued the follow-up with the FHS, performing weekly sessions of acupuncture and auriculotherapy, showing improvement in pain and emotional symptoms.

**DISCUSSION**

The patient was affected by the first symptoms at 29 years of age, therefore, in the second decade of her life. This differs from the literature, which indicates the fourth decade of life as the average age for the appearance of symptoms. Another point that can be evidenced in this case regarding epidemiology is the fact that it affects more women than men, and the patient under study is female\(^3\).
The complications described in the case report corresponded to the signs and symptoms evidenced in the patient’s daily life such as headache, asthenia, neck pain, paresthesia and spasms. In addition, she faced mainly physical limitations such as difficulty to walk, run and change her posture quickly; however, the patient believed she did not have major limitations due to the disease and was able to lead a normal life, working in and out of the house.

The clinical findings indicated in the present study differ from those in the literature. For example, a study conducted in an online community with 234 patients, most of them from the United States, reported that 45% of them had moderate or total disability due to pain and/or paresthesia, and 35% had moderate or total visual impairment. Although there were cognitive symptoms (thought and memory), they were less severe than physical symptoms, which are commonly associated with moderate to total disability.

Thus, the age at onset may be an important predictor of the location, extent, and clinical course of the disease. Cases with onset described after age 50 tend to develop severe early disability and myelitis, which may indicate that the spinal cord is more vulnerable to inflammation. In patients with onset before the age of 50, the optic nerves are more susceptible to inflammation. However, the number of relapses was lower in older patients in comparison to younger patients.

Despite the disease’s involvement in symptoms, the patient’s condition was not considered a serious prognosis. Thus, the shared care between PHC and other levels of care was observed, notwithstanding that at some moments the care was centered at the hospital level, with care technologies focused on medication and linked to medical professionals.

The PHC has different occupational categories which encompass multiple aspects around the individual so that they are taken care of in a comprehensive manner, providing care for the signs and symptoms resulting from the disease in the context in which they are inserted, and considered based on the everyday reality that surrounds them. Among the FHS professionals are: physicians, nurses, nursing technicians, dentists, and oral health technicians. At the Family Health Support Center (NASF – Núcleo de Apoio à Saúde da Família) psychologists, physiotherapists, physical education professionals, social workers, nutritionists, pharmacists and psychiatrists are available.

It was possible to verify that not all the tools available in the PHC were used. In this case the care could be accompanied by the NASF in addition to the FHS with the professionals that compose this nucleus, and who work together with the FHS with the objective of supporting, expanding, improving health care and care management, performing inter-sectoral and interdisciplinary actions, promoting, preventing and rehabilitating health.

Among the strategic areas of the NASF setting, mental health is encompassed as a focal point for the action of professionals in the nucleus; thus, the patient could have received psychological support, considering the feelings described by her from her perception of the disease, and also considering the propensity for depression resulting from illness, especially pain.

Psychological tests revealed signs of marked depression when compared to healthy controls for age and gender. Sensorial Quantitative Testing showed pronounced mechanical and thermal sensorial loss, strongly correlated with the pain in progress; these data emphasize the high prevalence of neuropathic pain and hyperalgesia in patients with Devic’s Disease.

Physiotherapy also could have contributed with exercises and physical therapy counseling to assist in the rehabilitation process and functional activity. Nursing itself, in addition to the resources exposed in this study, could have made use of complementary therapies for pain relief interventions, both in the outpatient clinic and in the hospital. Among the complementary therapies that could be used, the following stand out: application of cold or heat, control of the environment (comfort), distraction, teaching (procedure and treatment), stimulation of the imagination, stimulus to coping, massage, music therapy, relaxation, touch therapy and aromatherapy.

Moreover, other strategies could minimize the effects of pain, since it can affect quality of life, physical, psychological, social and spiritual dimensions of patients and their caregivers if not treated properly. Thus, non-pharmacological methods for pain relief are increasingly being used.

Pain has recently become to be considered as one of the main recognized symptoms of the disease, in addition to those already known. Thus, a study carried out with 11 patients with Devic’s Disease using the Quantitative Sensory Test showed that almost all (10 of the 11 patients) had neuropathic pain associated with the disease in the previous 3 months, and 8 of 11 patients indicated relevant pain at the time of the examination which can be related to alterations of the somatosensory function.

Although it was observed that the approaches used focused on drug therapy, it is positive that the team used Traditional Chinese Medicine through Integrative and Complementary Practices for managing the case. The National Policy on Integrative and Complementary Practices (PNPIC) defines the approach of these practices within the scope of the Unified Health System, taking into account the uniqueness of the human being and comprehensive care in health care as a form of preventive and therapeutic actions, minimizing the processes of illness.

In their conception of health and care, basic care physicians and nurses understand that the use of integrative practices for healthcare in Primary Care is related to an expanded understanding of health and disease, health promotion, physician–patient relationship, and self-care. For them, this technology of care is related to spiritual beliefs and practices, intuition and nutrition.

Integrative and Complementary Practices are gradually gaining greater visibility in the Brazilian context, although these are practices that have been carried out for a long time in other countries and cultures. Traditional Chinese Medicine, as the name suggests, originated in China thousands of years ago, encompassing a set of practices such as auriculotherapy, acupuncture, homeopathy, and phyotherapy, among others. It consists of an integrated system that...
seeks harmony and balance aiming at comprehensiveness through language that symbolically portrays the laws of nature. It is grounded in the yin-yang theory, and has the purpose of providing the means to balance this duality which is considered as two fundamental principles, interpreting all the phenomena in complementary opposites[10].

Auriculecterapy, one of the forms of care used by the PHC in the follow-up of this case, uses the stimulation of points in the auricular pavilion as a base for this practice. The points stimulated in the auricular pavilion correspond to points of the body such as organs, structures and functions. In this specific case, a set of points was used that corresponded to painful complaints resulting from the disease[10].

Acupuncture is a health intervention that can be used in isolation or associated with other therapeutic resources, addressing the health-disease process in human beings in a dynamic and comprehensive way. It is performed by precise stimulation of anatomical sites by the insertion of filiform needles for promoting, maintaining and recovering health[10].

Therefore, the treatment with ICP enabled a reduction in the medication such as carbamazepine (used for pain in the facial region) from 150 mg to 50 mg, since pain symptoms in the face decreased after beginning the treatment with acupuncture and auriculoterapia. In this sense, one study indicates a decrease in the use of analgesic drugs by cancer patients who use non-pharmacological therapies, among them complementary therapies for pain control[17].

These forms of care made it possible to follow the patient from a humanized perspective with a care plan decided in conjunction based on her complaints, and whose main symptom was pain. Thus, it is important to highlight the relevance of seeking possibilities of care available or innovative as alternative forms of care management that value the patient’s experience and pathology, with a focus on qualifying the care.

CONCLUSION

The care management process developed for a patient with Devic’s Disease involved the three levels of health care, highlighting the importance of multidisciplinary work between FHS/NASF and other levels of care. The PHC team coordinates care, promotes rehabilitation and avoids worsening of Devic’s Disease and other diseases, as recommended by the National Policy for Comprehensive Care for Persons with Rare Diseases.

The use of ICP has proven to be an effective strategy in the search for non-pharmacological, low-cost and effective ways of care, and according to the patient’s positive evaluation. Thus, the present study contributes to knowledge of professionals and people affected by demyelinating, inflammatory and degenerative diseases regarding referrals, rehabilitation and care possibilities at different levels of care.

As a limitation in this study we can point out the gap in the literature regarding the care management of patients with degenerative and demyelinating diseases in the PHC. Most studies on the subject focus on care at the hospital level. Thus, we highlight the need for further studies focusing on the use of complementary care therapies for patients with degenerative and demyelinating diseases.
REFERENCES


