



FREE THEME ARTICLE

NURSING CARE FOR PATIENTS CARRIERS OF WEST SYNDROME

CUIDADOS DE ENFERMAGEM A PACIENTES COM SÍNDROME DE WEST

CUIDADOS DE ENFERMERIA PARA PORTADORES DEL SÍNDROME DE WEST

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ABSTRACT

Objective: understand the syndrome under discussion for development of extended care in the area of pediatric nursing care intending to provide attention directed to children carriers of this syndrome. **Method:** it is a theoretical essay based on literature review, which allow the analyses of the published literature, printed and / or electronics articles magazines on a particular topic, in order to describe and discuss it under theoretical or contextual point of view. **Results:** at the end of the theoretical construction of West syndrome references, it's possible to notice that the implementation of Nursing Care System (NCS) was something essential to the mitigation and reduction of symptoms severity caused by the West Syndrome (WS). The nursing diagnostics which can be recognized in the WS case are: Impaired physical mobility; Risk of injury; chronic confusion, Risk of infection; Anxiety related to death. **Conclusion:** the NCS is considered a strategy of broad aspects when it comes to integral assistance for WS carriers. The NCS will be valuable for control and mitigation of severity caused by WS signs and symptoms. **Descriptors:** west syndrome; nursing; health care in integrals.

RESUMO

Objetivo: compreender a síndrome em debate para elaboração de cuidados ampliados na área da enfermagem pediátrica pretendendo fornecer uma atenção direcionada as crianças portadoras da doença. **Método:** trata-se de um ensaio teórico fundamentado em uma revisão bibliográfica do tipo narrativa, que permite a análise da literatura publicada, artigos de revista impressas e/ou eletrônicas sobre um determinado assunto, com o objetivo de descrevê-lo e discuti-lo, sob ponto de vista teórico ou contextual. **Resultados:** ao término da construção do referencial teórico da síndrome de West, pode-se perceber que a implementação da Sistematização da Assistência de Enfermagem (SAE) era algo imprescindível para a amenização e diminuição do agravo dos sintomas causados pela síndrome de West (SW). Os diagnósticos de enfermagem que podem ser adotados nos casos de SW são: Mobilidade Física Prejudicada; Risco para lesão; Confusão crônica; Risco para infecção; Ansiedade Relacionada à morte. **Conclusão:** a (SAE) é considerada uma estratégia de amplo aspecto quando se fala em assistência integral a esse paciente, e para o portador de SW, a SAE será de grande valia para controle e diminuição dos agravos gerados pelos sinais e sintomas. **Descritores:** síndrome de West; enfermagem; cuidados integrais em saúde.

RESUMEN

Objetivo: comprender el síndrome en cuestión para la elaboración de amplios cuidados en el campo de la enfermería pediátrica, pretendiendo proveer una atención direccionada a los niños portadores de la enfermedad. **Método:** se trata de un ensayo teórico basado en una revisión bibliográfica de tipo narrativo que permite el análisis de la literatura publicada, artículos de revista impresos y/o electrónicos sobre un determinado tema, con el objeto de describirlo y discutirlo, bajo el punto de vista teórico contextual. **Resultados:** al concluir la construcción del marco referencial teórico del Síndrome de West, se puede percibir que la implementación de la Sistematización de la Asistencia de Enfermería (SAE) era algo imprescindible para amenizar y reducir la severidad de los síntomas causados por el Síndrome de West (SW). Los diagnósticos de enfermería que pueden ser adoptados en los casos de SW son: Movilidad física perjudicada; Riesgo de lesión; Confusión crónica; Riesgo de infección; Ansiedad relacionada a la muerte. **Conclusión:** la SAE es considerada una estrategia de amplios aspectos cuando se habla de asistencia integral al portador del SW. La SAE será de gran valía para el control y reducción de la severidad causada por los signos y síntomas. **Descriptores:** síndrome de west, enfermería, cuidados integrales en salud.

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INTRODUÇÃO

The current research discusses the West syndrome, one among others child syndrome, in a perspective of integral and extended nursing assistance to the child carrier of this complex neuropathy.

The West Syndrome is characterized by spasm, *hyposrrhythmos* and retardation in the psychomotor development, though the patient could not present the all three symptoms at the same time.¹ The age of greatest incidence is between 3 to 12 months presenting spasms with short combinations of flexors and extensors movements of the head, torso and members².

Up against the symptoms, we aim to comprehend the syndrome in discussion to elaborate an extended health care in the pediatric nursery intending to provide a directed attention to the child carrier of this disease.

Therefore, this comprehension will make possible that children carriers of this deficiency receive the integral health care and promote a more dignified quality of life and advancement of pediatric knowledge for specific treatment with patients carriers of this disease and others similar that difficult or make impossible the normal development or growing of the psychic, motors, socials and biological patterns.

The interest in this subject arose from practical nursing classes of children and teen care, in the graduate course of nursery. In these classes the group of authors ran into a child diagnosed as carrier of the syndrome and, as they had difficulties to find in the literature the appropriate care guidance to apply to the case, they decided to plan the nursery care to this patient.

Thus, this study will permit to improve actions in the child health care field and in the pediatric nursing itself using strategies of rehabilitation, through a side by side work, join other professionals of different areas inserted in the care of the West Syndrome carrier, determining certain aspects of mayor risk related to this disease and that requires more care provided by carrying out decisions accordingly to the individual level of neuro-psychomotor and social compromise; examples of this are the emotional and psychological support to the family, making guidelines about possible complications and appropriate treatments concerning to the pathology.³

To have an understanding relating to nursing care, is necessary to have the application of systematization of nursing care (SAE) that in this pathology is very relevant to the delay of symptoms and support the family.

The study is of fundamental relevance to nursing due to the small area of literary production on the disease.

METODOLOGY

It is a theoretical essay based on a bibliographical review of narrative kind which allows the analysis of the published literature, magazine articles printed and / or electronically on a particular subject, in order to describe it and discuss it under theoretical or contextual point of view.⁴

The phases of the research were, respectively, the identification and location of the theoretical references to approach the topic in question, book report, the filing of the material found, obtaining of the pertinent information, and finally, the writing of the paper.⁴

We selected articles indexed in the databases Scientific Electronic Library Online (ScieELO), *Medical Literature Analysis and Retrieval System online* (MEDLINE), PUBMED and in the Nursing Data Bank (BDENF), using the following as search descriptors: “*Síndrome de West e plano de cuidados em enfermagem*”; “*Síndrome de West e plano de assistência em enfermagem*”; “*Síndrome de West e cuidados centrados no paciente*”; “*Síndrome de West e acesso aos cuidados de saúde*”; “*Síndrome de West e cuidado infantil*”.

In finding descriptors “*Síndrome de West e cuidados centrados no paciente*” (West Syndrome and patient-centered care); “*Síndrome de West e acesso aos cuidados de saúde*” (West Syndrome and Access to health care); “*Síndrome de West e cuidado infantil*” (West Syndrome and child care) were not results.

In the literature review we used the qualitative approach, since this allows you to enter deep into the essence of the theme. The publications were analyzed to verify the relationship between West Syndrome and nursing interventions, according to the study questions.⁴

RESULTS AND DISCUSSIONS

- Pathophysiological history of West syndrome

West Syndrome (WS) is an epileptic encephalopathy with nature characterized by spasms in flexion, hypsarrhythmia (characteristic EEG pattern), mental retardation associated with deficit or psychomotor decline, manifesting itself in individuals in the early stages of development, either in the intrauterine phase, from 2 to 4 month and before 2 years of age. It affects both sexes, but predominantly affects male children in all races.⁵

The disease was first identified in 1841 by English physician William James West in his own son, who performed the initial description of the problem, but only in 1951 is that Vasquez and Turner, Argentine doctors were able to define it more precisely with the advent of electroencephalography. They enjoined the diagnosis of infantile spasms with standard electroencephalogram identified as polymorphic cluttered and chaotic, calling him hypsarrhythmia⁶. Crises are translated by a jerking or spasms saved with the following features: sudden bending for the head with abduction and flexion of the upper legs (myoclonic spasms solid), it is common to issue a cry during the spasm.

Disease incidence is 0.25 and 0.60 cases for the equivalent of 1000 live births and its prevalence is 0.15 to 0.20 cases per 1000 live births in children under 11 years of life, being a 4000 or 6000 for each birth.³

Etymologically, it is presented as symptomatic (pre, peri and / or postnatal), cryptogenic and idiopathic.

As symptomatic, the disease has several definite causes for its occurrence divided in different periods of gestation, the prenatal period stand out as major cortical dysplasia (30%), hypoxic-ischemic event, as monosomies chromosomal abnormalities (syndrome Turner) and trisomy (Down syndrome), infections, syphilis, toxoplasmosis and rubella, and metabolic disorders, including phenylketonuria and neonatal adrenoleukodystrophy, perinatal, the most common causes are hypoglycemia and hypoxic-ischemic encephalopathy, and finally, in postnatal, SW comes intracranial hemorrhage due, trauma and infection by bacterial meningitis.⁷ Another fact that contributes to the emergence of the SW is the prematurity of the fetus.

In cryptogenic etiology the cause stems from organic abnormalities, without success in the neurological examination, finally, there isn't the idiopathic form a definition of an

underlying disease for the syndrome, including occurrence of normal psychomotor development.³

It is worth to remark that the causes may occur separately, grouped and included in the three periods of the fetus (prenatal, perinatal and postnatal).

Among the etiologies described the symptomatic has the worst prognosis and may evolve into Lennox-Gastaut syndrome, a severe form of childhood epilepsy syndrome⁸; cryptogenic and idiopathic forms show better response to treatment and reduction of disease manifestations resulting from but does not occur reverse at any time and may even result in death of the child carrier.³

The diagnosis is made through neurological examinations such as EEG, MRI, CT and neuroimaging, as well as clinical examination with emphasis on past history during the period of pregnancy, childbirth and neonatal, and physical examination. In addition, there are useful tests such as CBC, serology for syphilis as VDRL and metabolic demonstrative.³

Approaching forms of therapy, there are measures of surgical correction, as in cases of dysplasia, with drug vigabatina and employability of ACTH, aiming control of symptoms, and physical therapy, hydrotherapy and by motor-cognitive stimulation.^{2,5,3}

The physical therapy aims mainly to treat the sequelae acquired or minimize them as much as possible, reducing the deformities resulting from the disease through passive mobilization and stretching, including even performance of physiotherapy to strengthen the muscles of breathing due to hypotonia. Hydrotherapy is another feature highly effective, relieving spasticity heat provide by the pool that will enable joint range of motion, flexion and extension of the limbs.

• Nursing care plan

The nursing process is the focal point of the profession, being the regulator of nursing practice, showing a series of actions that must be developed before the patient. When applied the process results in a qualified approach of integral attention. We can define nursing process as

A methodological model that enables us to identify, understand, describe, explain and / or predict the human needs of individuals, families and communities in the face of life-cycle events or health problems, real or potential, and determine what aspects of these needs require a nursing professional

intervention.^{9:210}

The symptoms of the disease (epileptic spasm, mental and motor impairment) limit nursing care aimed at correcting the same, however, at the same time, it allows focusing on others priorities as routine cares.

A strong example is the administration of medications to control the disease that is usually prescribed vigabatrina and divalproex sodium, and drugs to control mainly the access crisis, stratagems and orientation of appropriate dosages, schedules and adverse effects arising from adequate help in the process of care and minimizing the effects of progressive syndrome.

To have applicability in a nursing process of methods used for the precatice is the Nursing Care System (NCS) which is divided into five important steps for the proper conduct of proceedings, they are: History, the collection of information to make certain changes; Diagnostic, interpretation of data; Planning, which is the strategy development; Implementation, which is the beginning and completion of the necessary actions to get results and Expected Results, which provide an indication of results achieved on the actions.¹⁰

For nursing provide the necessary and qualified assistance to the patient with West Syndrome and to the carrier family, it's necessary to perform the NCS.

● Nursing interventions for patients with the West Syndrome

Given the complexity of the syndrome in question and the need to plan assistance to these patients an effective tool for nursing is the identification of a history of pregnancy, identifying data gynecological, obstetrical, family and personal that might interfere in the normal course of pregnancy and contributing causal factors in the development of the West syndrome.

Similarly, monitoring of growth and normal development of the newborn permits to evaluate the degree of motor development, cognitive, affective and to identify others origin mechanisms of the disease in the postpartum, such as infections and bacterial meningitis.

Likewise, consistent guidance during the course of prenatal care, especially about certain diagnoses as VDRL tests to detect syphilis, TORCH serological group (cytomegalovirus, rubella and toxoplasmosis), complete blood count and HIV, can avert possible outbreaks etiological syndrome symptomatic in its constitution; to monitor weight and blood glucose using fasting glucose test and oral glucose tolerance also allow the recognition and prevent the development of hypoglycemia, the effective newborn screening postpartum allows, besides to detection of several other diseases, the presence of phenylketonuria, a metabolic disorder considered one of the causes of West Syndrome.¹¹

Speaking of that patient care is essential to involve the family. The moment of identification of an individual with this syndrome represents a hard impact on the family and changes the normal routine of its members; the psychological support means a great weapon of integral care at the moment of refusal and denial of family members, including parents, to a deficient newborn that requires extensive and almost exclusive dedication.

Emotional changes may occur to the family, with periods of anger, sadness, dismay and misunderstanding can lead to depression, both for intermittent periods. The support to the formal or informal caregiver is also part of the strategy for nursing care, and the family as the subject member of NCS, because care must be provided to the patient, family, environment and feelings that surround it.

The following table summarizes the plan of care developed by the authors of this text:

DIAGNOSTIC	PLANNING	IMPLEMENTATION	EVALUATION
Impaired physical Mobility Related to muscle atrophy evidenced by lack of movement	• Position in alignment to prevent complications such as pressure ulcers; • To promote the strengthening of the muscles	• Use footrest; • Avoid prolonged periods in one position; • Exercising body mechanics to increase muscle tone.	The patient affected by West Syndrome has strengthened the muscles and no complications related to impaired mobility.
Risk for injury Related to spasm, as evidences by abrupt movements	• To promote actions to prevent injuries related to seizures.	• Lateralize the patient's head to does not suffer asphyxia and injuries; • Move objects that offer risk, so they are far from the patient.	• The patient should not present injuries as resulting from spasms caused by the disease.
Chronic confusion, Related to mental deficit, evidenced by cognitive impairment and intellectual	• Slowing the progression of mental retardation;	• To provide medications that slow the progression of disability (by prescription); • Verbalize to the child even if no answer, to stimulate neural synapses, delaying the progression.	It should slow the progression of mental symptoms
Infection Risk Related to the amount of times that invasive procedures were made as evidenced by the need to submit to it.	• Conduct orientation to nursing staff	• Using aseptic technique during invasive procedures such as administering medication and bladder catheterization. • Verbalize about the correct technique, emphasizing the probable damage if not performed correctly.	The patient should not have complications arising from a breach of correct techniques.
Anxiety Linked to death evidenced by low life expectancy of the patient.	• Mitigate feelings of distress of the relatives of patients with the syndrome.	• Verbalizing about the evolution of the disease, so there is a preparation of future events that may happen. • Encourage social interaction of the family so there is an emotional support and distraction from them.	Family anxiety related to the thought of the possible patient's death will decrease.

Picture 1 constructed by authors

FINAL CONSIDERATIONS

West Syndrome is a little-known deficiency in the means of assistance and care, therefore, nursing care is poor in addressing the issues and individuals in their particulars. Thus, the development of a more defined and extensive attention to the carrier and family are priorities of the entire network of health professional, being the nursing process an example of intervention to improve the pathological and biopsychosocial conditions of the patient and his family.

It was understood that immediate care is the most recommended by experts in the range of care, but the nursing care systematization is considered a strategy of broad aspect when related integral assistance to this patient.

When dealing with major complications such as West syndrome, it's necessary an appropriate and effective clinical management to improve the quality of life of patients through care plans not only pharmacological, but that provide for the needs of comfort, psychological, among other, to the patient and the family.

Considering the constant need of trained human resources in health services, it's understandable that investing in continuing education and implementation of the NCS can be an essential tool for a quality nursing care to patients with this syndrome.

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