197 - GUILLAIN-BARRÉ SYNDROME: REVIEW OF LITERATURE

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INTRODUCTION

The Guillain-Barré Syndrome (GBS) is defined as an acute or subacute polyneuropathy and occurs in 60% of cases after some infectious disorder, being 50% of these after viral diseases. Typically, the viral infection comes before the motor infection in two to three weeks (CECATTO et al, 2003).

According to Tavares et al, (2000), the GBS is an autoimmune disease affecting the entire population at any time of year, affecting adults and children, men and women, regardless of social class and lifestyle, appearing to be more frequent with advancing age and more common in men, with an annual incidence in North America for two to four cases per 100,000 population, with similar epidemiological patterns worldwide.

Generally, the recovery process of patients suffering from this disease is slow, requiring long hospitalization, intensive care and multidisciplinary team that may be watching for any signs of instability in order to prevent complications.

In a three-month trial in a referral hospital in emergency rooms in Rio de Janeiro, was carried out a volunteer work with two young patients affected by GBS, and while every effort has been made, one of the cases had death as outcome. This fact has mobilized the nursing staff to reflect on this pathology, and especially the trainees involved, which, until then, had no knowledge of the pathophysiology and the nursing care provided to the carrier of this syndrome.

In this light, driven by the desire to gain more knowledge about this neurological and autoimmune disease, led to the literature search about the disease, especially with the scarcity of publications on the subject. Given this finding and knowing the importance of the presence of the nurse in assisting the holder of the GBS, as well as the smooth conduct of information and guidance given to their relatives, it was considered that these were motivating factors for this work.

In this sense, had the following objectives for this study: to review the literature on this syndrome emphasizing the clinical and complications that patients develop lifelong disease and to consider the conditioning factors and complications of patients affected with GBS.

MATERIAL AND METHODS

This is a literature review which aims to explain a problem from theoretical references to published documents, seeking to understand and analyze scientific contributions and cultural differences on a particular theme, not merely repeating what has been said or described, but allows the examination of a subject under a new approach, with innovative conclusions (MARCONI and LAKATOS, 2001).

For the development of this research, we sought to publications available in BIREME, specifically in the databases of the Literature of Latin American and Caribbean Health Sciences (LILACS) and International Literature in Health Sciences (Medline), the period was the years 1995 to 2008. The choice of these two databases was due to the fact that they are widely used by health professionals, promoting the multidisciplinary science, thus contributing to the construction of complex knowledge. The search was made through the junction of the descriptors "Guillain Barré Syndrome" and "Guillain Barré and Nursing", available in Descriptors in Health Sciences (DECS), a period from February to April 2008.

We extracted articles on this subject, a minority was related to nursing, in this case, only two, one national and one international, the rest were twenty for other professions, which represent the majority of our sample.

RESULTS

KNOWING THE GUILLAIN-BARRÉ SYNDROME

According to Grzesiuk and Santos (1999), the exact cause of GBS is still unknown. Noviello et al (2007) report that is autoimmune and some event happens after infection. And, although the pathogenic mechanisms are not yet fully established, it has been shown that the humoral and cell immune response contributes to its occurrence. In about 20% to 50% of cases can be detected autoantibodies anti-GM1 (anti-glycophospoholipid 1) and the process can be triggered by infection, the infectious agent most commonly listed as antecedent is Campylobacter jejuni.

It is a progressive peripheral neuropathy that affects the muscles of the human body, from the members and breathing, even the heart. It is usually characterized by weakness or paralysis that affects more than one member, generally symmetrical and associated with loss of tendon reflexes and increased protein concentration in cerebrospinal fluid (GOLDMAN, 2001).

For Funes, Montero and Carranza (2002 apud BENETTI and SILVA, 2006), is characterized by an acute inflammatory polyradiculoneuropathy of autoimmune origin and single phased. There are symmetrical progressive and ascending members paralyses and this may be presented with atypical forms and spontaneous remissions. The uptrend paralysis was first described more than a century ago, considering that Waldrop, in 1834, reported a case of a patient with probable GBS.

For Torres, Sánchez and Pérez (2003), the GBS is one of the most frequent forms of neuropathy, with more rapidly probable fatal evolution. To Quintero and Boza (1999), this syndrome has become the leading cause of flaccid paralysis after the eradication of polio.

According to Funes, Montero and Carranza (2002 apud BENETTI and SILVA, 2006) in the mid 50's, several pathological studies were carried out by French and German on the GBS. These studies revealed the presence of a mononuclear inflammatory infiltrate in nerves of patients who died, supported by Asbury and colleagues in 1969, from a series of autopsies of the dead with this pathology. In 1956, C. Miller Fisher discovered a variant of the syndrome characterized by total external ophthalmoplegia, severe ataxia and areflexia, variant that bears his name. In 1963, Melnick founds antibodies acting against nerve tissue in nineteen of thirty-eight patients with GBS. In 1964, this disease gets the name of Landry-Guillain-Barré-Strohl in honor of their researchers. Another variant that affected whether or not the myelin sheath of nerves, was described and named by Feasby and colleagues in 1986 of axonal GBS. Recently it has proposed the existence of another variant of inflammatory demyelinating polyradiculoneuropathy, this, predominantly, sensory.

According to Souza and Souza (2007), the dominant pathophysiological lesions of this syndrome result from multifocal infiltration of the myelin sheath by inflammatory mononuclear cells or the destruction of myelin antibody-mediated autoimmune

diseases. The individual with GBS produces antibodies against its own myelin (membrane of lipids and proteins that involve the nerves and facilitates the transmission of nerve stimulation) of the peripheral nerves and sometimes the proximal nerve roots and cranial nerves (nerves that emerge from a part of the brain called the brainstem and meets the specific functions of the head, neck and viscera). Generally, this inflammatory process is preceded by a severe infectious process that affects the synapse (union of nerve cells) that occurs between the motor root and peripheral nerves. As a result of inflammation, there is muscle involvement, leading to paralysis of these, muscle atonia, distal muscle strength deficit, loss of reflexes, decreased sensitivity of the skin in the lower leg and hands.

Lastra and Hedero (2002 apud BENETTI and SILVA, 2006) report as important diseases associated with the development of GBS: infection of upper respiratory tract, fever syndrome of unknown origin, gastrointestinal infections, diarrheal illness caused by Campylobacter jejuni; viral infectious diseases, especially those caused by cytomegalovirus, herpes virus, hepatitis A and B and human immunodeficiency virus, rabies vaccine and cancer.

Funes, Montero and Carranza (2002, apud BENETTI and SILVA, 2006) reported that GBS is a disease of worldwide distribution and its occurrence virtually independent of the season. It affects any age, but there are two peaks of higher incidence (young adults and elderly), and is a rare disease in children under one year of age. There is no clear predisposition for the acquisition of this syndrome by gender, but several studies found that men are more frequently affected than women. This condition, according to the authors, presents the annual incidence from 1.2 to 2.73 cases per 100,000 inhabitants and a greater incidence among the elderly population. In it, the annual incidence for the syndrome can reach 8.6 cases per 100,000 inhabitants over 70 years.

Platón et al (2003, apud BENETTI and SILVA, 2006) report that today, unlike what happened in ancient times, when mortality due to GBS was very high, only about 5% of patients die, due to a proper understanding of the development of the disease, which made possible better treatment.

Several studies, according to Tavares et al (2000) showed that most patients have elevated serum levels of tumor necrosis factor alpha (TNF-α), a cytokine highly toxic to the myelin sheath and Schuwan cell. This increase is directly correlated with the severity of the disease, since when the circulating levels of TNF-α are lower, there is clinical improvement in most patients.

Funes, Montero and Carranza (2002, apud BENETTI and SILVA, 2006) and Tavares et al (2000) support the idea that the GBS is related to an immune response that the body performs to infectious agents present in the body. Studies show that over 60% of patients with the syndrome suffered some type of infection in the weeks before the start. The infectious agent presents chemical molecules, which are recognized by the immune system as antigens. The body starts to produce specific response to these antigens leading to the production of specific antibodies. Chemical molecules present in the peripheral nerves and spinal roots may have structural similarity to the antigens and are attacked by antibodies produced according to the infection. The chemical similarity between these molecules is called molecular mimicry. These authors report that there are multiple infectious agents associated with the development of the syndrome and, in most cases, these agents are Unknown; it can now, however, be determined through epidemiological studies.

According to Goldman (2001), the symptoms are: tingling of the limbs, especially in the lower leg and hands, muscle atonia, decreased reflexes, decreased sensation, uncoordinated movements, muscle pain, blurred vision, difficulty breathing, dizziness, tachycardia, difficulty urinating or urinary incontinence, caused by inappropriate secretion of ADH (antidiuretic hormone), constipation, fainting, difficulty swallowing and distal strength deficits.

Platón et al. (2003, apud BENETTI and SILVA, 2006) argue that the GBS is typically characterized by a clinical triad that consists of paresthesias, general ascending weakness and areflexia, and is preceded in most cases for low back pain and muscle pain, fastly progressing with hypotonia, respiratory failure and dysautonomia, and the picture is complete in 80% of cases in the first three weeks, and recovery may take a few weeks or persist for up to six months; however, in up to 15% of cases, the standard clinical characteristic can not be observed due to the appearance of several atypical forms of the disease.

To Campellone (2004, apud BENETTI and SILVA, 2006), the manifestations of GBS rapidly progress (about a few days to weeks), causing muscle weakness or paralysis equally on both sides of the body, and weakness that starts in the legs, then extending to the arms. Is referred to as ascending paralysis, can occur at the same time tingling and pain in hands or feet. The initial phase of the disease is accompanied by rapid deterioration, more severe symptoms manifest themselves in a few hours and can last about three weeks, followed by a period of stability during which there is no change. Then it can show a phase of improvement/restoration, lasting a few days to six months, or even more. The author puts as typical symptoms: muscle weakness or paralysis, which started in the feet and legs, progressing to the arms and cranial nerves, with development that can occur within 24 to 72 hours. Also be seen as being typical symptoms: changes in sensations such as numbness, decreased sensitivity, tendon or muscle pain, which often accompanies or precedes the weakness. Other symptoms may be associated with disease: confused vision, difficulty in moving the muscles of face, palpitations and twitching, difficulty swallowing, difficulty breathing, temporary absence of respiratory movement, inability to perform movement and respiratory collapse.

Tavares et al (2000) also add that the weakness reaches its peak in the first two weeks of onset of disease, when then 20 to 30% of patients need the help of artificial ventilation, 40% are confined to bed, 20% need help walking, 10% could walk but not run and 10% keep only mild symptoms, with more than 90% of cases, loss of tendon reflexes. The autonomic nerves may also be compromised, leading many patients to submit an increase or sudden drops in blood pressure or pulse, excessive sweating, constipation, urinary retention, sinus tachycardia and bradycardia.

According to Brunner and Suddarth (2002), the GBS is considered a medical emergency, and the patient must be treated in an intensive care unit. The nature of the potentially fatal disease is related to difficulty in swallowing (aspiration may occur), stand-alone and deregulation, especially the potential for respiratory failure. The recovery of the patient is variable.

Noviello et al (2007) state that the diagnosis is based on the clinical features (progressive motor weakness and areflexia) and liquor analysis, which is the only laboratory criteria established. The elevation of the protein and the presence of ten or fewer mononuclear cells support the diagnosis. Electromyography helps differentiate the clinical and pathological characteristics of the disease.

Souza and Souza (2007) claim that about 95% of individuals with this disease survive and 75% recover completely. For some, the disease will continue for the rest of life. It can be fatal in some cases due to difficulty breathing. You can expect full recovery in patients in whom the symptoms disappear in three weeks. The need for mechanical ventilation and the lack of functional improvement in these three weeks after the disease had reached the peak signals are more severe.

The treatment of GBS consists primarily of supportive medical measures and in immunomodulation, according to Fonseca et al. (2004). For Torres, Sánchez and Pérez (2003), supportive measures of treatment are essential to avoid complications. These measures are: heparin to prevent pulmonary embolism, nutritional support to ensure immunocompetence and weaning from mechanical ventilation, chest physiotherapy to prevent atelectasis and pneumonia; general physiotherapy to avoid bodily contractures and psychological support to patients in order to achieve recovery.

The specific therapy currently used, according to Tavares et al. (2000), both techniques proved effective in treating the syndrome consisting of plasmapheresis and intravenous infusion of high doses of immunoglobulin. In plasmapheresis, the blood is removed, separating the plasma from blood cells. These are returned to the patient, achieving thus remove much of the antibodies and other circulating factors that probably have action in the pathogenesis of the syndrome. According Campellone (2004),

plasmapheresis can reduce the severity of symptoms and facilitate a faster recovery.

For Benetti and Silva (2006), treatment based on intravenous immunoglobulin has been considered the most appropriate choice and dose of 0.4g/kg/ day for five days. It leads to the achievement of results similar to those of plasma, however, achieve better functional level in a shorter time and with fewer side effects and limitations. The specific form of action of immunoglobulins is still unknown, however, there is evidence that they work by neutralizing antibodies that block the neuromuscular union by a dose-dependent mechanism mediated by antibodies.

To Quintero and Boza (1999), the use of corticosteroids may be considered, but is not established clear benefit to demonstrate the effectiveness of these drugs in the treatment of GBS. For Torres, Sanchez and Perez (2003), the use of intrathecal corticosteroids in betametazona dose of 0.8mg in alternate day for two weeks, has shown good results in young patients, however, this does not apply to aged 50 years, considering that in these, has been verified the occurrence of large amount of complications such as hyperglycemia, hypertension and gastrointestinal bleeding.

Fonseca et al. (2004) report that still experimental for the treatment of GBS, is an indication of b-interferon, there are other prospects for future treatment of this pathology, such as the use of growth factor nerve, of immunomodulatory cytokines and prevention, elimination and control of certain infections.

To Souza and Souza (2007), it should be done by a monitoring by health specialist. At the beginning of the disease, probably the user will need medical care during hospitalization, especially if you have difficulty breathing and swallowing involved in the acute setting; however, because the symptoms are decreasing, the disease can be monitored at home with the help of a nurse, and with periodic visits to the doctor.

FINAL CONSIDERATIONS

At the end of the study, we understand the importance of discussing health actions aimed at qualified and integrated practice, aimed at a none purely curative assistance, but holistic and reflective. It is important to clarify that the patient in no way should be exposed to embarrassing comments or be treated with differentiation. Its clinical course is developed with full support of professionals to promote the restoration of normal pattern of health.

The study emphasizes the importance of interaction between nurses and patient with GBS with family members and other professionals, identifying the need to study factors related to the hospital on the humanitarian perspective, and thereby acquire and enhance skills that support the planning and implementation of therapeutic nursing and to promote the minimization of the generating causes of stressful situations for the patient and companion.

Dealing with the sensitivity and individualized patient is something that requires professional patience, dedication, understanding and, above all, commitment.

The nurse must be aware and proficient in their trials and diagnostics, as in dealing with the uncertainties of human behavior, it is ultimately required of this professional stewardship in talking, acting, and plan its systematization and for these actions are carried out within the limits of the other and, consequently, their individuality.

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GUILLAIN-BARRÉ SYNDROME: REVIEW OF LITERATURE

ABSTRACT

GBS is an acute inflammatory demyelinating polyneuropathy of unknown etiology, characterized by flaccid paralysis affecting more than one member, usually symmetrical, with the absence of reflexes and increased liquor protein without pleocytosis. The symptoms are: tingling of the limbs, especially in the lower leg and hands, muscle atonia, decreased reflexes, decreased sensation, uncoordinated movements, muscle pain, blurred vision, difficulty breathing, dizziness, tachycardia, difficulty urination or urinary incontinence, caused by inappropriate secretion of ADH (antidiuretic hormone), constipation, fainting, difficulty swallowing and distal strength deficits. Had as objectives to research, conduct a literature review on the GBS highlighting the clinical and

complications that patients develop lifelong disease and to consider conditioning factors and complications to those afflicted with the syndrome. To develop the research, we aimed to publications available in BIREME, (Lilacs / Medline) between the years 1995 to 2008, using the keywords "Guillain Barré Syndrome" and "Guillain Barré and Nursing," from February to April 2008. GBS is a disease of worldwide distribution, occurring virtually independent of the season. It affects any age and there are two peaks of higher incidence (young adults and the elderly), is rare in children under one year old. There is no clear predisposition for the acquisition of this syndrome by gender, but several studies found that men are more frequently affected than women. The annual incidence for the syndrome can reach 8.6 cases per 100,000 inhabitants over 70 years. It is a relatively frequent disease, particularly compared to other neural diseases, being important the knowledge of it and the care of these patients who get ill, aiming to use in their professional practice of the Nursing Systematic Assistance.

KEYWORDS: Guillain-Barré Syndrome, Guillain-Barré and Nursing.

SYNDRÔME DE GUILLAIN BARRÉ: RÉVISION DE LITTÉRATURE

RÉSUMÉ

La SGB est une polyneuropathie démielisante inflammatoire aigüe, d´ éthiopatogénie inconnue, caracterisée par la paralisie flasque qui atteint plus d´un membre, en général symétrique, associée au manque de réflexes et augmentation de protéines au liquor, sans pléocytose. Les symptômes sont: sensation de fourmillement des membres, principalement dans la partie inférieure de la jambe et des mains, atonie musculaire, diminution des réflexes, diminution de la sensibilité, mouvements avec du manque de coordination, douleur musculaire, vision floue, difficulté pour respirer, vertige, taquicardie, difficulté pour uriner ou incontinence urinaire, entraînées par la sécretion inapropriée d´ADH (hormone antidiurétique), intestin arrêté, évanouissement, difficulté pour avaler et déficit de force distale. Il y a eu comme objectifs pour la recherche, réaliser une révision de littérature sur la SGB en détachant la clinique et les complications que les patients dévellopent au long de la maladie et considérer les facteurs conditionants et les complications aux atteints de la syndrome. Pour le dévellopement de la recherche, on a cherché des publications disponibles dans la Bireme, (Lilacs/Medline), entre les années 1995 à 2008, en utilisant les descripteurs “syndrome de Guillain Barré” et “syndrome de Guillain Barré et Infirmerie”, dans une période de février à avril 2008. La SGB est une pathologie de distribution mondiale, d´essor pratiquement indépendant de l´époque de l´année. Elle atteint n´importe quel âge, avec deux sommets de plus grande ocurrence (adultes jeunes et âgés), est rare entre les enfants mineurs d´un d´âge. Il n´y a pas de prédisposition claire pour l´aquisition de cette syndrome selon le sexe., mais plusieurs études ont observé que les hommes sont souvent plus atteints que les femmes. L´incidence annuelle peut arriver à 8,6 des cas pour chaque 100.000 habitants majeurs de 70 ans. C´est une maladie relativement fréquente, principalement si comparée à d´autres maladies neurales, étant important sa connaissance ainsi comme le soin avec ces patients qui en sont atteints, en envisageant l´utilisation dans la pratique professionnelle de l´Assistance en Infirmerie.

MOTS-CLÉS: Syndrome de Guillain Barré, Syndrome de Guillain Barré et Infirmerie.

SÍNDROME DE GUILLAIN BARRÉ: REVISIÓN DE LITERATURA

RESUMEN

A Síndrome de Guillain Barré (SGB) es una polineuropatia desmielinizante inflamatoria aguda, de etiopatogenia desconocida, caracterizada por parálisis flácida afectando más de un miembro, generalmente simétrica, asociada a la ausencia de reflejos y aumento de proteínas en el liquor, sin pleocitose. Los síntomas son: sensación de hormigueo de los miembros, principalmente en la porción inferior de la pierna y en las manos, atonía muscular, disminución de los reflejos, disminución de la sensibilidad, movimientos sin coordinación, dolor muscular, visión empañada, dificultad para respirar, vértigo, taquicardia, dificultad para orinar o incontinencia urinaria, causados por la secreción inapropiada de ADH (hormona que disminuye la secreción de la orina), intestino arrestado, desmayo, dificultad para engullir y déficit de fuerza periférica. Se tuvo como objetivos para la pesquisa, realizar una revisión de literatura acerca de la SGB resaltando la clínica y las complicaciones que los pacientes desarrollan a lo largo de la enfermedad y considerar los factores condicionantes y complicaciones a los atacados con el síndrome. Para el desarrollo de la pesquisa, se buscó publicaciones disponibles en la Bireme, (Lilacs/Medline), entre los años de 1995 a 2008, utilizándose los descriptores “síndrome de Guillain Barré” y “síndrome de Guillain Barré y Enfermería”, en un período de febrero a abril de 2008. A SGB es una patología de distribución mundial, de ocurrencia prácticamente independiente de la época del año. Ataca cualquier edad, existiendo dos picos de mayor ocurrencia (adultos jóvenes y de edad), es rara en niños menores de un año de edad. No hay predisposición clara para adquisición de ese síndrome en cuanto al sexo, pero varios estudios observaron que los hombres son frecuentemente más afectados que las mujeres. La incidencia anual para el síndrome puede llegar hasta 8,6 casos para cada 100.000 habitantes mayores de 70 años. Es una enfermedad relativamente frecuente, principalmente se comparada a otras enfermedades del sistema nervioso, siendo importante el conocimiento de la misma bien como el cuidado con estos pacientes que la adquiere, visando a la utilización en su práctica profesional de la Sistematización de la Asistencia en Enfermería.

PALABRAS CLAVE: Síndrome de Guillain Barré, Síndrome de Guillain Barré y Enfermería.

SÍNDROME DE GUILLAIN BARRÉ: REVISÃO DE LITERATURA

RESUMO

A SGB é uma polineuropatia desmielinizante inflamatória aguda, de etiopatogenia desconhecida, caracterizada por paralisia flácida afetando mais de um membro, geralmente simétrica, associada à ausência de reflexos e aumento de proteínas no liquor, sem pleocitose. Os sintomas são: sensação de formigamento dos membros, principalmente na porção inferior da perna e nas mãos, atonia muscular, diminuição dos reflexos, diminuição da sensibilidade, movimentos incoordenados, dor muscular, visão embaçada, dificuldade para respirar, tontura, taquicardia, dificuldade para urinar ou incontinência urinária, causados pela secreção inapropriada de ADH (hormônio antidiurético), intestino preso, desmaio, dificuldade para engolir e déficit de força distal. Teve-se como objetivos para a pesquisa, realizar uma revisão de literatura acerca da SGB ressaltando a clínica e as complicações que os pacientes desenvolvem ao longo da doença e considerar os fatores condicionantes e complicações aos acometidos com a síndrome. Para o desenvolvimento da pesquisa, buscou-se publicações disponíveis na Bireme, (Lilacs/Medline), entre os anos de 1995 a 2008, utilizando-se os descritores “síndrome de Guillain Barré” e “síndrome de Guillain Barré e Enfermagem”, num período de fevereiro a abril de 2008. A SGB é uma patologia de distribuição mundial, de ocorrência praticamente independe da época do ano. Acomete qualquer idade, existindo dois picos de maior ocorrência (adultos jovens e idosos), é rara em crianças menores de um ano de idade. Não há predisposição clara para aquisição dessa síndrome quanto ao sexo, mas vários estudos observaram que os homens são freqüentemente mais afetados que as mulheres. A incidência anual para a síndrome pode chegar até 8,6 casos para cada 100.000 habitantes maiores de 70 anos. É uma doença relativamente freqüente, principalmente se comparada a outras doenças neurais, sendo importante o conhecimento da mesma bem como o cuidado com estes pacientes que a adquire, visando à utilização na sua prática profissional da Sistematização da Assistência em Enfermagem.

PALAVRAS CHAVE: Síndrome de Guillain Barré, Síndrome de Guillain Barré e Enfermagem.

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