Review Article

A Review Study on Chronic Inflammatory Demyelinating Polyneuropathy

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Abstract

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare immune-mediated disease in which an abnormal immune response causes demyelination and damage to the axons of peripheral nerves. This study is a systematic review of the literature. The screening of the abstracts was performed on the abstrackrBeta web-based software platform. The etiology, pathogenesis, modern methods of CIDP treatment, as well as the economic consequences of the spread of the disease, reflected in the scientific literature, are considered. Forty five 45 complete texts and nineteen conference procedures were found on the study of disease transmission n 9, humanistic burden n 7, current treatment n 40, and financial burden n 8 of CIDP. Epidemiological study have appeared the frequency and predominance of 0.2 1.6 and 0.8 8.9 per 100,000, depending on topography and symptomatic criteria. Six main types of therapy have been reported in publications on modern treatment methods: intravenous immunoglobulins, subcutaneous immunoglobulins, corticosteroids, plasmapheresis, immunosuppressants, and immunomodulators. According to the analyzed data, indications for the choice between these methods of treatment, as well as dosage regimens and frequency of use, have not been determined. Thus, CIDP is an autoimmune disease that requires constant monitoring and correction of therapy if necessary.

Keywords: Polyneuropathy, Plasmapheresis, Nerve conduction, Autoimmune disease

Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare immune-mediated disease in which an abnormal immune response causes demyelination and damage to the axons of peripheral nerves [1]. The exact etiology of CIDP remains unknown. The new data suggest a possible genetic contribution to CIDP. In a recent study, patients with CIDP had a high frequency of perforin gene variations that disrupt the function of cytotoxic T cells and natural killer cells [2, 3]. Patients have progressive weakness, impaired sensitivity in the legs and arms, loss of deep tendon reflexes (areflexia), and fatigue [4]. CIDP is a long-term condition with a variable course, which can be recurrent-remitting, stepwise-progressive, or gradually progressive [5]. Axon damage occurs with further progression of the disease, leading to worsening symptoms [6]. The severity of the disease for the patient can be assessed using several functional outcomes, which are mainly focused on functional disorders, disability, and impaired ability to perform everyday actions [7].

Tools for assessing disorders include the scale of causes and treatment of inflammatory neuropathy (INCAT) and the scale of general disability constructed by Rush (I-RODS) [8].

Guidelines of the European Federation of Neurological Societies/Peripheral Nerve Societies (EFNS/PNS) contain recommendations for treating CIDP to reduce symptoms and, if possible, maintain long-term remission [9]. Recommendations for long-term management are not provided due to lack of evidence. In addition, treatment may be associated with a decrease in the patient's independence.

Treatment of CIDP is complicated by diagnostic difficulties [10, 11]. The disease can manifest itself in various symptoms, and at least 15 diagnostic criteria describe CIDP and its variants [5, 12]. Various CIDP manifestations and misinterpreting nerve conduction studies lead to a high frequency of misdiagnoses [4]. This may lead to inaccurate treatment, as patients with CIDP may be misdiagnosed with other polyneuropathies, such as anti myelin associated

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glycoprotein neuropathy MAG or polyneuropathy of Lyrics disorder polyneuropathy, organomegaly, endocrinopathy, M protein, and skin changes, which require other treatment [13].

To date, no publication provides a comprehensive overview of the burden of CIDP in terms of epidemiology, modern treatment methods, and economic costs [14, 15].

Objective: to conduct a systematic review of the available data on chronic inflammatory demyelinating polyneuropathy from the point of view of epidemiology, modern treatment methods, and economic costs.

Materials and Methods

A review of scientific literature in the WoS and Scopus systems, research articles in the MEDLINE (including In-process), and Embase databases was conducted, and a review of scientific papers was carried out. The screening of the abstracts was performed on the abstrackrBeta web-based software platform. The etiology, pathogenesis, modern methods of CIDP treatment, as well as the economic consequences of the spread of the disease, reflected in the scientific literature, are considered.

Results and Discussion

Forty five 45 complete texts and 19 conference materials were found on the study of disease transmission n 9, humanistic burden n 7, current treatment n 40, and financial burden n 8 of CIDP. Epidemiological thinks about have appeared the frequency and predominance of 0.2 1.6 and 0.8 8.9 per 100,000, depending on geology and symptomatic criteria. Six fundamental sorts of treatment have been detailed in distributions on present day treatment strategies intravenous immunoglobulins, subcutaneous immunoglobulins, corticosteroids, plasmapheresis, immunosuppressants, and immunomodulators. Treatment can be burdensome due to side impacts and decreased freedom caused by the treatment conditions. In Germany, the UK, France, and the USA, the financial complexity of CIDP was due to the coordinate costs of treatment and hospitalization.

Clinical Picture, Etiology, and Pathogenesis of the Disease

First described almost 50 years ago, chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare autoimmune disease characterized by progressive peripheral neuropathy. The exact etiology of CIDP remains unknown. The clinical picture of CIDP is expressed in the form of symmetrical sensorimotor polyneuropathy with a monotonous, gradual course. Patients have progressive weakness, impaired sensitivity in the legs and arms, loss of deep tendon reflexes (areflexia), and fatigue [6]. This disease is a long-term condition with a variable course, which can be recurrent-remitting, stepwise-progressive, or gradually progressive [5]. The variety of manifestations makes it possible to distinguish the classical form of CIDP and atypical types **(Table 1)**.

Table 1. Clinical manifestations of different forms of CIDP

CIDP Form	Clinical manifestations
Classic	There is a symmetrical muscular hypotension in all parts of the limbs, a violation of sensitivity, which increases for more than 2 months. The development of the disease is slow, monotonous, and gradual and may be accompanied by exacerbations.
Distal	Pronounced asymmetric lesions of the hands, forearms, hands, shins
Focal	There is a lesion of one or more nerve fibers in the brachial or lumbosacral plexus.
Isolated (sensitive)	The process of demyelination affects only sensory nerve fibers

Epidemiological studies have confirmed that CIDP is a rare disease (incidence: 0,2–1,6/100 000 people per year, prevalence: 0.8–10.3 per 100,000 people) [16]. The data on morbidity and prevalence differed, probably due to differences in the study sample size (n = 19-360) and diagnostic criteria.

According to the data, from 2021 to 2022, about 200 cases of neurological diseases were registered in Russia. Of these, CIDPS increases after 12 months **(Table 2)**.

Table 2. Disability due to CIDP

Years	Due to CIDP	Disability (number)	% of Disability
2021	7	157	3.4
2022	14	236	3.9

The diagnosis of CIDP is established after a detailed study of the patient's anamnesis, complaints, and the specifics of the symptoms of the disease, data from electroneuromyography, magnetic resonance imaging of the spine, and ultrasound of nerve fibers. CIDP is difficult to diagnose, but early diagnosis can be crucial to prevent irreversible nerve damage.

Features of Treatment of Chronic Inflammatory Demyelinating Polyneuropathy

With CIDP, cellular and humoral components of the immune system attack the myelin of large peripheral nerve fibers, which leads to demyelination, manifested by weakness, numbness, paresthesia, and sensory ataxia. As the disease progresses, axon loss occurs secondary to demyelination and is associated with unfavorable prognoses [17]. No action has been identified as a biomarker for CIDP in general. Hence, specific autoantibodies have been identified against paranodal proteins in Ranvier interceptions in peripheral nerves in about 10% of patients, neurofascin-186, gliomedin, and contactin-1. However, some may have prognostic value and predict a poor response to specific immunomodulatory drugs [18, 19].

Initial treatment options include corticosteroids, intravenous immunoglobulin, and therapeutic plasmapheresis. Subcutaneous administration of immunoglobulin represents a new opportunity for patients with CIDP, which can increase independence and improve tolerance. At the same time, treatment with corticosteroid pharmaceuticals should be carried out with strict monitoring of blood pressure, blood sugar, cholesterol, and calcium preparations. Bone density control is carried out by densitometry. Immunosuppressive agents can become a worthy alternative to corticosteroids. Replacement of drugs is necessary in the presence of side effects from taking steroids and low effectiveness of treatment due to the inability to reduce the therapeutic dose [20].

Six main types of therapy have been reported in publications on modern treatment methods: intravenous immunoglobulins, subcutaneous immunoglobulins, corticosteroids, plasmapheresis, immunosuppressants, and immunomodulators. Treatment can be burdensome due to side effects and reduced independence caused by the treatment conditions. In Germany, Great Britain, France, and the USA, the economic complexity of CIDP was due to the direct costs of treatment and hospitalization [21].

This review presents the first systematic assessment of the burden of CIDP disease. According to the analysis of the data obtained, we found that CIDP is a rare disease associated with a violation of the quality of life, especially regarding the physical well-being of the patient and the economic costs of his treatment. Epidemiological studies have confirmed that CIDP is a rare disease (incidence: 0,2–1,6/100 000 people per year, prevalence: 0.8–10.3 per 100,000 people). In the population, the condition is more often manifested in women **(Figure 1)**. At the same time, accurate data on the incidence and prevalence of CIDP in the world is difficult to analyze due to the disparity of data, as well as a large gap in the sample size of subjects in studies aimed at studying the disease (n = 19-360). In addition, the lack of uniform diagnostic criteria for CIDP makes it difficult to search and analyze scientific data on this disease. Small sample sizes are common when identifying and recruiting patients with rare diseases [22].

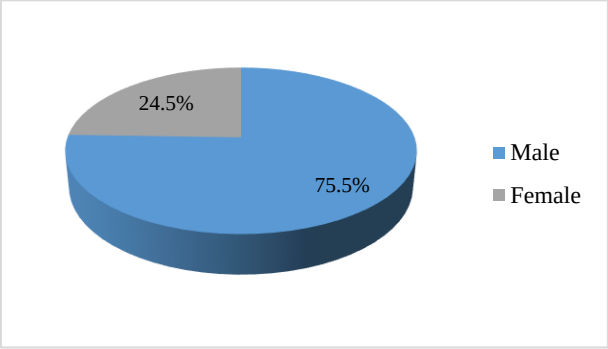

Figure 1. Distribution of morbidity in the population by gender

There is no data in the scientific literature on the study of individual clinical characteristics of false-positive and false-negative diagnoses of CIDP, which requires further investigation. It is also necessary to study the dynamics of patients' physical and mental well-being with different forms of CIDP.

Modern treatment methods can improve patients' well-being but are also associated with tolerance problems and problems around the route of administration [20]. Both intravenous immunoglobulins and corticosteroids are effective in achieving a response to treatment. Following the recommendations of EFNS/PNS, it can be used as a first-line therapy [23, 24]. However, long-term use of corticosteroids is associated with serious side effects in patients, such as hypertension, kushingoid appearance, and gastrointestinal complaints [25-27].

Conclusion

Epidemiological studies have shown the incidence and prevalence of 0.2-1.6 and 0.8-8.9 per 100,000, depending on geography and diagnostic criteria. Six main types of therapy have been reported in publications on modern treatment methods: intravenous immunoglobulins, subcutaneous immunoglobulins, corticosteroids, plasmapheresis, immunosuppressants, and immunomodulators. Treatment can be burdensome due to side effects and reduced independence caused by the treatment conditions. In Germany, the UK, France, and the USA, the economic complexity of CIDP was due to the direct costs of treatment and hospitalization.

According to the analyzed data, indications for the choice between these methods of treatment, as well as dosage regimens and frequency of use, have not been determined. With the ineffectiveness of the first line of therapy - glucocorticosteroids and intravenous immunoglobulins, the effectiveness of which has been proven by several multicenter randomized placebo-controlled studies, it is impossible to use other immunomodulating agents, the use of which is not regulated, since no studies have been conducted to prove their effectiveness. Thus, CIDP is an autoimmune disease that requires constant monitoring and correction of therapy if necessary.

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