Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is a neurological condition characterised by a gradual development of muscle weakness, loss of feeling and changes in sensation in the arms and legs. CIDP is generally viewed as the chronic form of Guillain-Barré Syndrome (GBS), an acute, post infectious paralysing disorder, but can be differentiated from it by clinical course, prognosis and response to certain treatments [1]. CIDP occurs when the immune system mistakenly attacks the peripheral nerves-the nerves that lie outside the brain and spinal cord. Instead of defending against an infection the immune system attacks part of the nerve damaging it so that it can no longer function leaving the patient with various neurological symptoms. During this immune attack the fatty layer of the nerve fiber, the myelin, which functions like an insulating layer on a cable becomes stripped away leaving the nerve fibre bare and unable to conduct electrical impulses efficiently. The symptoms of CIDP develop for at least two months after onset and continue to evolve for months or even years. The cause of this aberrant immune attack is unknown. It is a relatively rare disorder with a prevalence of between 2 to7 per 100,000[2] adults, can start at any age and can affect either sex but is more common in males.

Symptoms and clinical course

Each person with CIDP experiences different symptoms with varying degrees of severity. Initial sensory symptoms usually include pins and needles, a burning sensation or numbness in the fingers and toes. Initial muscle weakness is often reported as a leaden or wooden feeling in the arms or legs and can also present as difficulty in using the hands to grip objects. There is usually a loss of reflexes such as the knee or ankle jerk. Although symptoms can remain relatively mild in some patients, in the majority they progress to severe disability with difficulty walking or paralysis of arms and legs. CIDP can follow either a progressive course where the symptoms become worse over time or a relapsing and remitting course where the symptoms improve only to come back again months or even years later in a manner similar to Multiple Sclerosis.

Diagnosis

Diagnosis of CIDP is based upon the clinical features, nerve conduction studies and laboratory tests. Nerve conduction studies are performed to test the function of the nerve and to determine whether the myelin has been damaged or removed. A demyelinated nerve fibre is unable to conduct electrical signals as fast or as efficiently as a healthy nerve fibre. Further laboratory tests such as a spinal tap/lumber puncture to analyse cerebrospinal fluid or a nerve biopsy can be carried out to support a diagnosis of CIDP or rule to out other conditions .MRI studies may show enlargement or inflammation of nerve.

Treatments

There are a number of therapies currently used to treat CIDP and although 70-90% [3] of patients respond to one of them most require long-term therapy and up to 30% respond to none of the recommended treatments. Successful treatments improve strength and sensation and the ability to perform daily tasks which has great impact on the quality of life. It is important to begin treatment early in the disease course to prevent further nerve damage which can lead to permanent disability. Hence early diagnosis is essential. Due to different responses to therapy in each patient treatment regimens must be individualised.

Most of the current treatments work by suppressing the immune system which stops the attack on the nerve and relieves symptoms. Such treatments carry the risk of suppressing the normal immune response to infection and patients therefore need to be closely monitored.

Corticosteroids

Corticosteroids such as prednisone are used as a treatment in CIDP due to their anti-inflammatory effects and have been shown to be beneficial in clinical trials. However, the many adverse side effects associated with long term steroid use such as obesity, hypertension, diabetes and osteoporosis warrant the development of alternative treatments.

Intravenous immunoglobulin (IVIg)

IVIg is a blood product that contains the pooled antibodies (immunoglobulin G) from over one thousand healthy people. IVIg exerts a number of effects on the immune system that could result in resolution of inflammation but the precise mechanisms of action are still being investigated. IVIg is administered intravenously over a 2-5 day period at intervals of 3-4 weeks. The improvement in disability after treatment with IVIg lasts from 2-6 weeks so further doses and intervals need to be determined on a case by case basis. IVIg is expensive but has become the preferred treatment,because of its safety and modest side effects. However in much of the developing world it cannot be afforded.

Plasmapheresis or Plasma Exchange

Plasmapheresis or plasma exchange is the removal of the fluid (plasma) from the blood. The patient's blood is pumped through a machine that separates the blood cells from the plasma and replaces the patient's plasma with albumin. Initial treatment requires several plasma exchanges over a two week period followed by an exchange every 6-8 weeks to maintain the effects. This process removes harmful antibodies or immune factors that are present in the circulation and may contribute to the nerve damage. Plasmapharesis has a beneficial short term effect as an initial therapy in two thirds of patients but is usually combined with other treatments for stabilisation and maintenance [4].

Immunosuppressive drugs

A wide range of immunosuppressive drugs has been used in the treatment of CIDP with varying success and none has been proven to be effective in a controlled clinical trial. Immunosuppressive drugs are generally used as second line treatments if the patient has not responded to steroids, IVIg or PE. Many of these drugs reduce the number of immune cells in the blood so patients must be monitored with regular blood tests throughout the treatment.

Conclusions

Due to medical research considerable progress has been made in the development of effective treatments that improve the quality of life for many patients and allow them to live normal lives. However, more research is needed to identify the cause and precise mechanisms of the disease process which will lead to more directed and affordable treatments with fewer side effects. In the most populous regions of the world's the best treatment, IVIg cannot be afforded.

References

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