# Facts About
# Plasmapheresis


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# Plasmapheresis and Autoimmune Disease

Many diseases, including myasthenia gravis, Lambert-Eaton syndrome, Guillain-Barré syndrome and others, are caused by a so-called autoimmune, or self-immune, process. In autoimmune conditions, the body's immune system mistakenly turns against itself, attacking its own tissues. Some of the specialized cells involved in this process can attack tissues directly, while others can produce substances known as antibodies that circulate in the blood and carry out the attack. Antibodies produced against the body's own tissues are known as autoantibodies.

Dr. Peter Dau helped develop plasmapheresis in the 1970s with MDA support. At that time, he was at Children's Hospital of San Francisco. He's now at Evanston Hospital in Evanston, Ill.

Treatment with medications that suppress the activities of the immune system and/or reduce inflammation of tissues has been the most common approach to autoimmune disease for more than 30 years. Many new immunosuppressants have become available since the 1960s, but all the medications used to treat autoimmune disease have serious side effects when taken in high doses for months or years.

In the 1970s, with the support of the Muscular Dystrophy Association, researchers developed a new approach to the treatment of autoimmune conditions. Instead of trying to change the immune system with medication alone, they thought it might be possible to mechanically remove autoantibodies from the bloodstream in a process similar to that used in an "artificial kidney," or dialysis, treatment. The procedure became known as plasmapheresis, meaning plasma separation. It's also known as plasma exchange.

Medications that suppress the immune system or reduce inflammation often are combined with plasmapheresis, but they usually can be given in lower doses than when used alone.

Today, plasmapheresis is widely accepted for the treatment of myasthenia gravis, Lambert-Eaton syndrome, Guillain-Barré syndrome and chronic demyelinating polyneuropathy. Its effectiveness in other conditions, such as multiple sclerosis, polymyositis and dermatomyositis, is not as well-established.

## What is plasmapheresis?

Plasmapheresis is a process in which the fluid part of the blood, called plasma, is removed from blood cells by a device known as a cell separator. The separator works either by passing the blood at high speed to separate the cells from the fluid or by passing the blood through a membrane with pores so small that only the fluid part of the blood can pass through. The cells are returned to the person undergoing treatment, while the plasma, which contains the antibodies, is discarded and replaced with other fluids. Medication to keep the blood from clotting (an anticoagulant) is given through a vein during the procedure.

## What's involved in a plasmapheresis treatment?

A plasmapheresis treatment takes several hours and can be done on an outpatient basis. It can be uncomfortable but is normally not painful. The number of treatments needed varies greatly depending on the particular disease and the person's general condition. An average course of plasma exchange is six to 10 treatments over two to 10 weeks. In some centers, treatments are performed once a week, while in others, more than one weekly treatment is done.

A person undergoing plasmapheresis can lie in a bed or sit in a reclining chair. A small, thin tube (catheter) is placed in a

large vein, usually the one in the crook of the arm, and another tube is placed in the opposite hand or foot (so that at least one arm can move freely during the procedure). Blood is taken to the separator from one tube, while the separated blood cells, combined with replacement fluids, are returned to the patient through the other tube.

The amount of blood outside the body at any one time is much less than the amount ordinarily donated in a blood bank.

## Are there risks associated with plasmapheresis?

Yes, but most can be controlled. Any unusual symptoms should be immediately reported to the doctor or the person in charge of the procedure. Symptoms that may seem trivial sometimes herald the onset of a serious complication.

The most common problem is a drop in blood pressure, which can be experienced as faintness, dizziness, blurred vision, coldness, sweating or abdominal cramps. A drop in blood pressure is remedied by lowering the patient's head, raising the legs and giving intravenous fluid.

Bleeding can occasionally occur because of the medications used to keep the blood from clotting during the procedure. Some of these medications can cause other adverse reactions, which begin with tingling around the mouth or in the limbs, muscle cramps or a metallic taste in the mouth. If allowed to progress, these reactions can lead to an irregular heartbeat or seizures.

An allergic reaction to the solutions used to replace the plasma or to the sterilizing agents used for the tubing can be a true emergency. This type of reaction usually begins with itching, wheezing or a rash. The plasma exchange must be stopped and the person treated with intravenous medications.

Excessive suppression of the immune system can temporarily occur with plasmapheresis, since the procedure isn't selective about which antibodies it removes. In time, the body can replenish its supply of needed antibodies, but some physicians give these intravenously after each plasmapheresis treatment. Outpatients may have to take special precautions against infection.

Medication dosages need careful observation and adjustment in people being treated with plasmapheresis because some drugs can be removed from the blood or changed by the procedure.

## How long does it take to see improvement?

Improvement sometimes can occur within days, especially in myasthenia gravis. In other conditions, especially where there is extensive tissue damage, improvement is slower but still can occur within weeks.

## Does MDA pay for plasmapheresis?

MDA supported pioneering research to develop plasmapheresis. However, payment for this procedure is not among the many services included in MDA's program. A number of health insurance plans do cover the procedure.

## Where are plasmapheresis treatments offered?

Plasmapheresis is performed at many major medical centers around the country. MDA clinic directors can offer advice about the availability of this treatment and its use for specific conditions.

# MDA's Purpose and Programs

The Muscular Dystrophy Association fights neuromuscular diseases through an unparalleled worldwide research effort. The following diseases are included in MDA's program:

## Muscular Dystrophies

Myotonic dystrophy *(Steinert disease)*
Duchenne muscular dystrophy
Becker muscular dystrophy
Limb-girdle muscular dystrophy
Facioscapulohumeral muscular dystrophy
Congenital muscular dystrophy
Oculopharyngeal muscular dystrophy
Distal muscular dystrophy
Emery-Dreifuss muscular dystrophy

## Motor Neuron Diseases

Amyotrophic lateral sclerosis *(ALS)*
Infantile progressive spinal muscular atrophy *(Type 1, Werdnig-Hoffmann disease)*
Intermediate spinal muscular atrophy *(Type 2)*
Juvenile spinal muscular atrophy *(Type 3, Kugelberg-Welander disease)*
Adult spinal muscular atrophy *(Type 4)*
Spinal-bulbar muscular atrophy (*Kennedy disease)*

## Inflammatory Myopathies

Polymyositis
Dermatomyositis
Inclusion-body myositis

## Diseases of Neuromuscular Junction

Myasthenia gravis
Lambert-Eaton (myasthenic) syndrome
Congenital myasthenic syndromes

## Diseases of Peripheral Nerve

Charcot-Marie-Tooth disease
Friedreich's ataxia
Dejerine-Sottas disease

## Metabolic Diseases of Muscle

Phosphorylase deficiency *(McArdle disease)*
Acid maltase deficiency *(Pompe disease)*
Phosphofructokinase deficiency *(Tarui disease)*
Debrancher enzyme deficiency *(Cori or Forbes disease)*
Mitochondrial myopathy
Carnitine deficiency
Carnitine palmityl transferase deficiency
Phosphoglycerate kinase deficiency
Phosphoglycerate mutase deficiency
Lactate dehydrogenase deficiency
Myoadenylate deaminase deficiency

## Myopathies Due to Endocrine Abnormalities

Hyperthyroid myopathy
Hypothyroid myopathy

## Other Myopathies

Myotonia congenita
Paramyotonia congenita
Central core disease
Nemaline myopathy
Myotubular myopathy
Periodic paralysis

MDA Muscular Dystrophy Association
Fighting Muscle Disease

MDA's website, mda.org, is constantly updated with the latest research news and information about the diseases in its program. Follow MDA on Facebook, Twitter and YouTube.

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